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Revision topics in otology

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About this book

This book has been written with exam going students in mind. An average student finds it really difficult to revise all topics prior to examination. There is also the added disadvantage of time management. They have to juggle with preparations for different subjects. This book has been structured in such a way that frequently asked questions in the examination have been dealt with in an objective manner. Exam going student just by reading this book will be able to rapidly revise the important topics.

This e book is definitely not a replacement for standard text books in the subject. This book is not meant to supplement the standard literature in the speciality and help the students during their exam preparations.

About the author

Dr Balasubramanian T is currently Professor of Otolaryngology at Stanley Medical College Chennai India. He has extensive experience in teaching under graduate and post graduate students of otolaryngology. He has been engaged in web based teaching for the past 6 years. He is running websites for the benefit of otolaryngology students. These sites are free to access.

1. www.drtbalu.com
2. www.drtbalu.co.in
3. www.otolaryngology.co.in
4. www.drtbalu.net
5. <http://atlas.otolaryngology.co.in/>
6. www.rhinology.in
7. www.drtbalu.wikidot.com
8. <http://ebooks.otolaryngology.co.in/>
9. <http://entslides.wikidot.com/>
10. www.jorl.net (online journal of otolaryngology)

Students are welcome to visit these sites.

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52. [Presbyacusis](#)
53. [Otitis externa](#)
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58. [Glomus jugulare](#)
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61. [Puretone audiometry](#)
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Deaf Mutism

Deaf-mutism ¹ is generally due to a loss of hearing before the age of 2 - 3 years which hinders learning to speak. Most cases result from acute infectious diseases such as measles, epidemic meningitis, encephalitis, typhoid, otitis media, toxic effects of drugs, etc. Congenital deaf-mutism ² is also a possible etiology. Endocrine causes like Pendred syndrome involves deafness along with the presence of goitre. The child's intelligence is normal.

For normal speech to develop in a child, the hearing sensation should be intact. The first three years of life is considered to be crucial for the development of speech. Loss of hearing due to any cause during this crucial period of development causes the development of speech to falter.

Congenitally deaf child is also a mute. In congenital deafness there is failure of normal development of cochlea, leading on to deformities i.e. Mondini defect etc. Congenitally deaf and mute child can be identified by the absence of oculo vestibular reflex. This is the reflex that makes the eye deviate to opposite side when the head is bent to one side. In children with acquired deaf mutism this reflex is intact.

Management:

Rehabilitation is the only way. In the past these children were taught to communicate using sign language. Now days the children, if they have residual hearing should be provided with hearing amplification devices (hearing aids). They should also undergo a rigorous course of speech therapy.

The recent advances in the field of cochlear implant have added another useful dimension in the management of these patients.

References:

1. Ladd, P. (2003). *Understanding Deaf Culture*. In Search of Deaf hood. Toronto: Multilingual Matters
2. Aycock, W. L. and Ingalls, T. H. (1946): Amer. J. fed. Sci., 212, 366.
3. <https://sites.google.com/site/drtbalusotolaryngology/otology/deaf-mutism>

Endolymphatic Hydrops

Endolymphatic hydrops is also known as Meniere's disease(Thiagarajan) . Classically described by Prosper Meniere ¹ during 19th century, this condition is due to swelling up of the endolymphatic sac. This causes fluctuating low tone sensory neural hearing loss, tinnitus and episodic vertigo.

Causes:

1. Idiopathic (Most common)
2. Viral infections
3. Allergy
4. Syphilis
5. Cochlear otosclerosis
6. Hormonal imbalances causing water retention

Pathophysiology:

Meniere's disease by definition is idiopathic endolymphatic hydrops characterised by roaring tinnitus, vertigo, fluctuating hearing loss. Even though sometimes erroneously used interchangeably, Meniere's disease is different from endolymphatic hydrops. It should be borne in mind that the term endolymphatic hydrops indicates the underlying pathophysiological mechanism of Meniere's disease. Endolymphatic hydrops can in fact be classified as primary and secondary according to the causative factors involved. Primary endolymphatic hydrops is in fact the classic Meniere's disease where the underlying etiology is unknown. In secondary hydrops the etiopathogenesis of the underlying disorder is clearly elucidable.

Etiology and pathophysiology of Meniere's disease is still unclear. Most commonly accepted theory being over distension of the membranous labyrinth due to excessive endolymphatic fluid volume. This accumulation of endolymph may be caused by impaired absorption of endolymph in the endolymphatic duct and sac, or excessive secretion of endolymph. These hypotheses are based on demonstrable presence of endolymphatic hydrops in patients with signs and symptoms of Meniere's disease. It has also been demonstrated that these patients have reduced vascularization and fibrosis of perisaccular tissue causing a reduction in the

absorptive capacity of the endolymphatic sac. Experimentally also it has been clearly demonstrated that obliteration of endolymphatic sac will induce endolymphatic hydrops.

Studies have clearly demonstrated that early increase in the volume of endolymphatic fluid (relative to the perilymphatic compartment) occurs in the pars inferior portion of the membranous labyrinth. This portion includes cochlear duct and saccule. In advanced stages of the disease the whole of the membranous labyrinth can be involved. Cochlear hydrops was seen in all patients of Meniere's disease and saccular hydrops was seen in most. Utricular hydrops was rarely seen.

The degree of endolymphatic space expansion is highly variable. The endolymphatic space bulged in the region of helicotrema(Thiagarajan) in half of the cases, while saccule bulged against the foot plate in 60% of cases, into a semicircular canal usually horizontal in 1/3 of cases. Fibrous adhesions can form between the saccule and the under surface of the Stapedial foot plate. This contact may explain Hennebert's sign (subjective vertigo, tonic eye deviation and nystagmus observed during a pressure induced excursion of the foot plate). It may also explain the "Tullio phenomenon" which is experienced by some Meniere's patients.

Diagnosis:

Is made by performing Glycerol test. Glycerol in doses of 1 - 2ml /Kg is given to the patient in lemon water. If the patient shows improvement from the symptoms and hearing then the diagnosis is confirmed.

Pure tone audiometry:

Classically demonstrates low frequency Sensorineural hearing loss.

Management:

1. Bed rest
2. Labyrinthine sedatives
3. Diuretics
4. Salt free diet
5. Endolymphatic sac decompression surgery in resistant cases

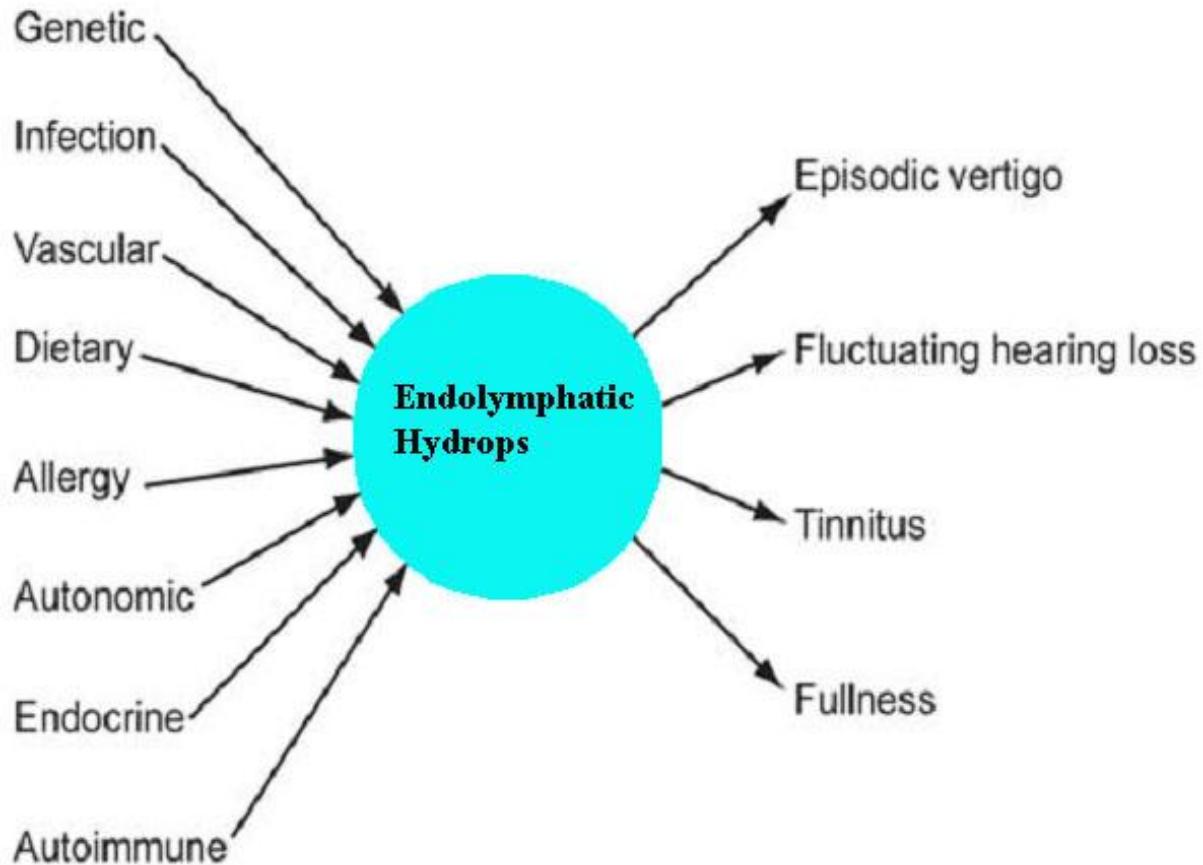


Diagram illustrating causes and manifestations of endolymphatic hydrops

References:

1. <http://www.drtbalu.co.in/news/article.php?story=20110308105233591>
2. <http://ebooks.otolaryngology.co.in/node/12>

Thiagarajan, B. Ménière's disease, Geetha R.

Furuncle external auditory canal ¹

It is also known as acute localised otitis externa / circumscribed otitis externa. This is a localised infection usually found to involve the lateral 1/3 of the external canal. It also has a propensity to involve the posterior superior aspect of the external canal. This is caused due to obstruction of the apopilosebaceous units found extensively in this area.

Etiology: ²

Trauma to skin in this area followed by infection is commonly attributed cause. The organism responsible is commonly staph aureus.

Symptoms:

1. Localised pain
2. Localised itching
3. Purulent discharge if the abscess ruptures
4. If oedema or abscess occludes the external canal hearing loss can occur.

Signs:

1. Erythema of the skin
2. Localised abscess formation

Management:

If the abscess is pointing it can be treated by incision and drainage. Oral antibiotics should be used. The preferred drug of choice is penicillin of first generation cephalosporins. Anti-inflammatory drugs can be used to reduce inflammation and pain.

These patients must be advised to cut their nails short and to keep their hands clean, since this is the commonest route of infection.

References:

1. <https://sites.google.com/site/drtbalusotolaryngology/otology/furuncle-external-auditory-canal>
2. http://www.drtbalu.co.in/otitis_ext.html

Otitis externa ²

Synonyms: Infections of the external ear

Definition: Otitis externa is defined as infection / inflammation of the external auditory canal / auricle. It can range in its severity between mild infections to a more severe one. It is one of the most common diseases involving the external ear.

Classification:

Otitis externa is classified as follows:

1. Acute diffuse otitis externa (commonly caused by bacteria)
2. Acute localised otitis externa (commonly furuncle)
3. Chronic otitis externa
4. Eczematous otitis externa
5. Fungal otitis externa
6. Malignant otitis externa

Predisposing factors responsible for otitis externa:

Under normal conditions the skin lining the external auditory canal is well protected by its self-cleansing mechanism. In diseased conditions several factors may come into play in the pathogenesis of otitis externa.

1. Absence of cerumen: The cerumen plays an important role in the protection of the external canal. It protects the external canal from moisture. It also has anti-bacterial properties which help in the protection of the external canal. The cerumen also lowers the pH of the external canal making it difficult for the bacterial pathogens to colonize.
2. Removal of cerumen by ear buds: is one of the common causes of otitis externa. The act of removal traumatises the skin lining of the external canal making it vulnerable to infections.
3. Frequent exposure to water: external canal when constantly bathed in water loses its ability to protect itself. The presence of water macerates the skin lining of the external canal and also increases the pH of the external canal making it more favourable for bacterial colonisation. This condition is common in swimmers.

Acute diffuse otitis externa:

This is also known as the swimmers ear. This is an inflammatory condition involving the external canal in a diffuse manner. This condition is common in swimmers because of the propensity for the external canal to be exposed to water for long durations. This exposure leads to maceration of the external canal skin, and also lowers the pH of the external canal providing an environment favourable to infections.

Main symptoms:

1. Itching in the external canal
2. Tenderness on palpation
3. Aural fullness rarely occur due to the reduction in size of the external canal lumen due to oedema
4. Rarely stenosis of the external canal may occur causing accumulation of debris and secretions

Common signs:

1. Erythema of the external canal
2. Oedema of external canal
3. Secretions from the external canal (weeping canal)
4. Pain on mastication
5. Pulling of helix in a postero superior direction cause pain
6. In advanced cases fever and lymphadenopathy may occur (pre and post auricular nodes may be involved)

Stages of acute diffuse otitis externa: (Senturia)¹

Preinflammatory stage: is characterised by intense itching, edema and sensation of fullness in the ear.

Inflammatory stage: may be divided into mild, moderate and severe.

Mild acute inflammatory stage: here the cardinal features are increased itching, pain, mild erythema and oedema of the external canal skin. At later stages exfoliation of skin with minimal amount of cloudy secretions may be seen in the external canal.

Moderate acute inflammatory stage: in this stage the itching and tenderness of the external canal intensifies. The external canal is narrowed due to oedema and accumulation of epithelial debris.

Severe acute inflammatory type: In this stage pain becomes intolerable to such an extent the patient may refuse to eat, the lumen of the external canal becomes totally obliterated due to oedema and accumulated epithelial debris. Otorrhoea may become purulent. In addition regional nodes may also be involved. Infections from the external canal may involve the parotid gland via the fissures of santorini.

Common organisms involved: *Pseudomonas aeruginosa* and *staphylococcus aureus* are commonly cultured from the external canal of these patients. The normal commensals like *staphylococcus epidermidis* and *corynebacteria* are conspicuously absent.

Management:

The aim is two fold:

1. Resolving the infection
2. Promoting the external canal skin's recovery to its original state.

Firstly the canal is cleaned atraumatically by gentle suctioning and debridement under microscope. Topical hydrogen peroxide solution instilled will help the process of debridement.

A cotton wick dipped in I.G. paint can be inserted in to the external canal and allowed to stay for a day. This will reduce the external canal skin oedema and will increase the size of the meatus. Ear drops containing a mixture of neomycin and 1% hydrocortisone may be instilled as ear drops at least three times a day. In addition to the antibiotic and anti-inflammatory effects this drug reduces the pH of the external canal making it more resistant to the organisms.

In severe cases oral antibiotics and anti-inflammatory drugs can be resorted to. Quinolones are commonly used oral antibiotic.

Acute localised otitis externa: This condition is otherwise known as furunculosis or circumscribed otitis externa. This is a localised infection usually found to involve the lateral 1/3 of the external canal. It also has a propensity to involve the posterior superior aspect of the external canal. This is caused due to obstruction of the apopilosebaceous units found extensively in this area.

Trauma to skin in this area followed by infection is commonly attributed cause. The organism responsible is commonly staph aureus.

Symptoms:

1. Localised pain
2. Localised itching
3. Purulent discharge if the abscess ruptures
4. If oedema or abscess occludes the external canal hearing loss can occur.

Signs:

1. Erythema of the skin
2. Localised abscess formation

Management:

If the abscess is pointing it can be treated by incision and drainage. Oral antibiotics should be used. The preferred drug of choice is penicillin of first generation cephalosporins. Anti-inflammatory drugs can be used to reduce inflammation and pain.

These patients must be advised to cut their nails short and to keep their hands clean, since this is the commonest route of infection.

Chronic otitis externa:

This is a chronic infection / inflammation involving the skin lining of the external canal. There is thickening of the skin lining of the external canal due to persistent low grade infection / inflammation.

Symptoms:

1. Unrelenting pruritus
2. Mild pain
3. Presence of dry skin in the external canal

Signs:

1. Asteatosis (lack of cerumen)
2. Hypertrophic external canal skin
3. Presence of dry flaky skin in the external canal
4. Mild tenderness on ear manipulation
5. Rarely muco purulent otorrhoea

Cultures from the external canal of these patients are highly unreliable because they would have been using various antibiotic drops to surmount the problem.

Management:

This involves extensive use of acetic acid ear drops. This helps to reduce the pH of the skin lining the external canal making it more resistant to bacterial infections. In intractable cases steroid drops can be tried. Antibiotic drops may not be useful in these patients.

Surgery is indicated in extreme cases. A canalplasty is performed to widen the external canal. The involved skin may be removed to be replaced by a split thickness graft.

Eczematous otitis externa:

This condition includes various dermatologic conditions involving the skin of the external canal. It may range from atopic dermatitis, contact dermatitis, seborrheic dermatitis, neuro dermatitis, infantile eczema etc.

This condition is characterised by intense itching, in fact this could be the only complaint of the patient. On examination, erythema of the external canal skin may be seen. There may also be associated scaling and oozing from the canal skin.

Success lies in the management of the underlying dermatologic condition.

Otomycosis:

It is also known as Fungal otitis externa. This is the commonest type of otitis externa in tropical countries. This condition is associated with increased ear canal moisture, or following treatment of otitis externa by prolonged use of topical antibiotics. The protective cerumen layer is absent in these patients. This condition is more common in diabetics.

Symptoms:

1. Intense itching
2. Pain when otitis externa is coexistent
3. Blocking sensation due to the presence of fungal ball

Signs:

1. Inflamed external canal skin
2. External canal tenderness
3. Fungal debris (black in case of aspergillus and white in the case of candida). Invariably the infection is mixed type.

Management:

The condition is managed by careful aural toileting to remove the fungal balls. The best way to remove fungus from the ear canal is by aural syringing. Antifungal ear drops of clotrimazole can be administered. If secondary infections are present oral antibiotics and anti-inflammatory drugs may be resorted to.

References:

1. Senturia B.H. 1956 Evaluation of factors which may be of importance in the production of external ear infections Journal of investigative dermatology 27 291-315
2. http://www.drtbalu.co.in/otitis_ext.html

Cholesteatoma ¹

Definition of cholesteatoma: Cholesteatoma is defined as a cystic bag like structure lined by stratified squamous epithelium on a fibrous matrix. This sac contains desquamated squamous epithelium. This sac is present in the attic region. Cholesteatoma is also defined as 'skin in wrong place'. Cholesteatoma is known to contain all the layers of skin epithelium. The basal layer (germinating layer) is present on the outer surface of cholesteatoma sac in contact with the walls of the middle ear cleft.

Theories of bone invasion by cholesteatoma:

1. Pressure theory - states that increase in the pressure caused by enlarging cholesteatoma cause bone erosion. Ischemia has been attributed as the cause in this theory.
2. Enzymatic theory: Inside the cholesteatoma are present multinucleated osteoclasts and histiocytes. These cells release acid phosphatase, collagenase and other proteolytic enzymes. These enzymes are known to cause bone erosion.
3. Pyogenic osteitis: Pyogenic bacteria may release enzymes which could cause bone desorption.

Types of cholesteatoma:

1. Congenital cholesteatoma
2. Primary acquired cholesteatoma
3. Secondary acquired cholesteatoma

Congenital cholesteatoma: is known to arise from embryonic cell rests present in the middle ear cavity and temporal bone. These cell rests are known to commonly occur in cerebello pontine angle and petrous apex. In fact congenital cholesteatoma is seen as a whitish mass behind an intact tympanic membrane.

Derlacki and Clemis laid down the following as criteria to diagnose congenital cholesteatoma:

1. The patient should not have previous episodes of middle ear disease
2. Ear drum must be intact and normal

3. It is purely an incidental finding

4. If discharge and ear drum perforation is present then it should be construed that congenital cholesteatoma has managed to erode the tympanic membrane.

Clinical features: The disorder is an incidental finding. The common location of congenital cholesteatoma is the antero superior quadrant of tympanic membrane, postero superior quadrant being the next common site of involvement. Anteriorly situated congenital cholesteatomas are known to affect the Eustachian tube function causing conductive deafness due to middle ear effusion; whereas posterior congenital cholesteatoma is known to cause conductive deafness due to impairment of ossicular chain mobility.

Staging of congenital cholesteatoma:

Staging as suggested by Derlacki and Clemis: They were the first to stage congenital cholesteatoma. They classified congenital cholesteatoma into

1. Petrous pyramid cholesteatoma
2. Cholesteatoma involving the mastoid cavity
3. Cholesteatoma involving the middle ear cavity.

Potsic suggested the following staging mechanism:

Stage I: Single quadrant involvement with no ossicular / mastoid involvement.

Stage II: Multiple quadrant involvement with no ossicular / mastoid involvement

Stage III: Ossicular involvement without mastoid involvement

Stage IV: Mastoid extension

Nelson's staging:

Type I: Involvement of mesotympanum without involvement of incus / stapes

Type II: Involvement of mesotympanum / attic along with erosion of ossicles without extension into the mastoid cavity

Type III: Involvement of mesotympanum with mastoid extension

Staging this disease will help in deciding the modality of treatment and in predicting the long term prognosis.

Acquired Cholesteatoma:

This can be divided into two types, primary acquired and secondary acquired cholesteatomas.

Primary acquired cholesteatoma: In this condition there is no history of pre-existing or previous episodes of otitis media or perforation. Lesions just arise from the attic region of the middle ear.

Secondary acquired cholesteatoma: always follows active middle ear infection which manages to destroy the ear drum along with the annulus. This type of destruction is common in acute necrotising otitis media following exanthematous fevers like measles etc.

Theories to explain pathogenesis of cholesteatoma:

Various theories have been postulated to explain the pathogenesis of cholesteatoma. They are:

1. Cawthrone theory: This theory suggested by cawthrone in 1963 suggested that cholesteatoma always originated from congenital embryonic cell rests present in various areas of the temporal bone.
2. Theory of immigration: This theory was suggested by Tumarkin. He was of the view that cholesteatoma was derived by immigration of squamous epithelium from the deep portion of the external auditory canal into the middle ear cleft through a marginal perforation or a total perforation of the ear drum as seen in acute necrotising otitis media.
3. Theory of invagination: This theory was suggested by Toss. He theorised that persistent negative pressure in the attic region causes invagination of pars flaccida causing a retraction pocket. This retraction pocket becomes later filled with desquamated epithelial debris which forms a nidus for the infection to occur later. Common organisms known to infect this keratin debris are Psuedomonas, E. coli, B. Proteus etc.

Toss also classified attic retraction pockets into 4 grades:

1. Grade I: The retracted pars flaccida is not in contact with the neck of the malleus.
2. Grade II: The retracted pars flaccida is in contact with the neck of the malleus to such an extent that it seems to clothe the neck of the malleus.
3. Grade III: Here in addition to the retracted pars flaccida being in contact with the neck of the malleus there is also a limited erosion of the outer attic wall or scutum.

4. Grade IV: In this grade in addition to all the above said changes there is severe erosion of the outer attic wall or scutum.

4. Metaplastic theory: This theory was first suggested by Wendt in 1873. He took into consideration the histological changes seen in various portions of the middle ear cavity. The attic area of the middle ear cavity is lined by pavement type of epithelium. This epithelium undergoes metaplastic changes in response to subclinical infection. This metaplastic mucosa is squamous in nature thereby forming a nidus for cholesteatoma formation in the attic region.

Of all the above mentioned theories, the theory of invagination appears to be the most plausible one currently explaining the various pathologic features of cholesteatoma.

Clinical features of acquired cholesteatoma:

Ear discharge: is scanty and foul smelling. In fact the odour is best described as musty in nature. This is due to the presence of saprophytic infection and osteitis.

Hearing loss: is commonly conductive in nature. Some patients may even surprisingly have a normal hearing despite the presence of a huge cholesteatoma. This normal hearing could be attributed to the bridging effects of cholesteatomatous mass.

Sensorineural hearing loss if present could be attributed to the absorption of toxins through the round window membrane, or may be due to use of ototoxic antibiotics topically on a long term basis.

Ear ache: if present could be attributed to the presence of co-existing otitis externa, or presence of extradural abscess.

Tinnitus if present may indicate imminent sensorineural hearing loss.

Vertigo may be present if there is erosion of lateral semicircular canal by the cholesteatomatous matrix. Fistula test if performed is positive in these patients.

Fistula test: This test is positive if there is a third window is present in the labyrinth due to the erosion of the labyrinthine bone. This commonly occurs in the lateral semicircular canal area. This test is performed using a snugly fitting Siegle's pneumatic speculum and slowly applying pressure by compressing the pneumatic bulb. If labyrinthine fistula is present the patient will feel giddy and will have nystagmus.

Facial palsy may indicate erosion of facial nerve canal with involvement of facial nerve.

On examination:

There is destruction of the outer attic wall, with presence of attic perforation. Cholesteatomatous flakes may be seen through the perforation like cotton wool.

There is associated sagging of the posterior superior meatal wall.

Hearing tests indicate conductive deafness commonly if labyrinth is uninvolved. It may turn out to be sensorineural hearing loss if there is associated erosion of the labyrinth.

X ray mastoids may show sclerosis with presence of cavity.

Management:

Since this is a surgical problem modified radical mastoidectomy is advocated in almost all of these patients.

The aims of the surgical procedure are as follows:

1. To exteriorise the disease
2. To create adequate ventilation to the middle ear cavity
3. To create a permanent skin lined cavity exposed to the exterior.

References:

1. <https://sites.google.com/site/drtbalusotolaryngology/otology/cholesteatoma>
2. <http://ebooks.otolaryngology.co.in/node/28>

Bell's palsy ⁴

Bell's palsy is defined as idiopathic lower motor neuron type of facial nerve paralysis. This is in fact the most common type of facial palsy. This condition was first described by Sir Charles Bell one century ago.

This condition is mostly unilateral, and rarely bilateral. Bell's palsy is a diagnosis of exclusion, which must be made only after excluding all the known causes of facial nerve paralysis.

Pathophysiology: Etiology and pathophysiology is highly controversial. The patient gives history of going to bed normally, and waking up with facial palsy, or there is a history of bus / train travel with the patient seated close to the window. Bell's palsy is a diagnosis of exclusion. All known causes of facial paralysis should be excluded before this diagnosis is entertained.⁵

1. Exposure to cold air has been postulated as one of the causes
2. Viral infections involving the nerve sheath.

There is inflammation of the facial nerve causing it to swell up. Since it is enclosed inside a rigid bony canal it has virtually no space to expand causing the damage to the nerve. The labyrinthine segment of the facial canal is the narrowest portion of the whole facial canal (about 0.6mm).

Clinical features:

The patient wakes up with lower motor neuron type of facial paralysis.

1. Inability to close the ipsilateral eye
2. Reduction of tearing in the ipsilateral eye
3. Deviation of the angle of the mouth to the opposite side
4. Drooling of saliva
5. Metallic taste in the tongue
6. Inability to wrinkle the forehead
7. Bell's phenomenon (rolling of eyeball upwards)

8. Hyperacusis since nerve to stapedius muscle is a branch of facial nerve.

This condition is very rare in pregnant women, and if present it tends to be very severe with poor recovery.

Prognosis is excellent. 99% of patients recovering completely.

Management:

1. Eye care: The patient should wear glasses to protect cornea. (Black glasses are preferable), use of artificial tears.
2. Regular physiotherapy (Balloon blowing)
3. Cheek / eye massage
4. Steroids ^{2,3}: Very useful in early stages of the disease
5. Antiviral drugs like acyclovir ¹ have been tried with varying degrees of success
6. Facial nerve decompression can be considered in patients who don't show signs of recovery within 6 months

References:

1. Sullivan FM, Swan IR, Donnan PT et al. (October 2007). "Early treatment with prednisolone or acyclovir in Bell's palsy". *N. Engl. J. Med.* **357** (16): 1598–607
2. Salinas RA, Alvarez G, Daly F, Ferreira J (2010). Salinas, Rodrigo A. ed. "Corticosteroids for Bell's palsy (idiopathic facial paralysis)". *Cochrane Database Syst Rev* **3** (3): CD001942.
3. Lockhart P, Daly F, Pitkethly M, Comerford N, Sullivan F (2009). Lockhart, Pauline. ed. "Antiviral treatment for Bell's palsy (idiopathic facial paralysis)". *Cochrane Database Syst Rev* (4): CD001869. [doi:10.1002/14651858.CD001869.pub4](https://doi.org/10.1002/14651858.CD001869.pub4). PMID [19821283](https://pubmed.ncbi.nlm.nih.gov/19821283/)
4. <https://sites.google.com/site/drtbalusotolaryngology/otology/bells-palsy>
5. Ahmed A (2005). "When is facial paralysis Bell palsy? Current diagnosis and treatment". *Cleve Clin J Med* **72**(5): 398–401, 405. [doi:10.3949/ccjm.72.5.398](https://doi.org/10.3949/ccjm.72.5.398). PMID [15929453](https://pubmed.ncbi.nlm.nih.gov/15929453/)

Otomycosis ²

Otomycosis: is superficial mycotic infection of skin lining the external auditory canal. This commonly occurs in humid seasons.

Otomycosis can be classified into primary and secondary otomycosis.

Primary otomycosis: Commonly occurs during humid conditions when there is excessive moisture content in the atmosphere. This excessive moisture makes the external canal skin soggy and oedematous. This predisposes to fungal infections.

Secondary otomycosis: Occurs in immunocompromised individuals and in persons who have preexisting CSOM. Patients with CSOM usually apply broad spectrum antibiotic ear drops. This ear drops not only kills pathogens but also the natural commensals causing secondary fungal infections.

Common fungal species involved: ¹

1. *Candida albicans*
2. *Candida tropicalis*
3. *Aspergillus niger*
4. *Aspergillus fumigatus*.

Of these fungi candida infections cause whitish wet plaques within the ear canal. The plaques may also appear leathery. The aspergillus niger appear as black plaques in the external auditory canal.

Clinical features:

1. Intense itching of external canal
2. Inflammation and scaling of external canal skin
3. White / black plaques seen depending on the type of fungal infection
4. Intense pain in the ear

Predisposing factors for Otomycosis include:

1. Humid climate
2. Presence of cerumen
3. Instrumentation of the ear

4. Immunocompromised host
5. Indiscriminate use of topical antibiotic & steroid ear drops

Treatment:

1. Dry mopping to remove plaques
2. Antifungal ear drops
3. Anti inflammatory drugs in case of acute inflammation

References:

1. Kaur R, Mittal N, Kakkar M, Aggarwal AK, Mathur MD (August 2000). "Otomycosis: a clinicomycologic study". *Ear Nose Throat J* **79** (8): 606–9. [PMID 10969470](#)
2. <https://sites.google.com/site/drtbalusotolaryngology/otology/otomycosis>

Tuning fork tests

Introduction:

These tests are performed in order to subjectively assess a person's hearing acuity. This test can in fact be performed by using tuning forks of the following frequencies (254 Hz, 512 Hz, and 1024 Hz). Frequencies below 254 Hz are better felt than heard and hence are not used. Sensitivity for frequencies above 1024 Hz is rather poor and hence is not used.

Prerequisites for an ideal tuning fork:

1. It should be made of a good alloy
2. It should vibrate at the specified frequency
3. It should be capable of maintaining the vibration for one full minute
4. It should not produce any overtones

Methodology of using tuning fork:

The tuning fork must be struck against a firm surface (rubber pad / elbow of the examiner). The fork should be struck at the junction of upper 1/3 and lower 2/3 of the fork. It is this area of the fork which is capable of maximum vibration.

The vibrating fork should be held parallel to the acoustic axis of the ear being tested.

Advantages of tuning fork tests:

1. Easy to perform
2. Can even be performed at bed side
3. Will give a rough estimate of the patient's hearing acuity

The following tests can be performed using a tuning fork:

1. Rinne test

2. Weber test
3. ABC test
4. Bing test
5. Politzer test
6. Bing Entotic test
7. Stenger's test
8. Gelle test
9. Chimani-Moos test

Rinne test:

Rinne's test: is a tuning fork test used to clinically test hearing deficiencies in patients. It is designed to compare air conduction with bone conduction thresholds. Under normal circumstances, air conduction is better than bone conduction.

Ideally 512 tuning fork is used. It should be struck against the elbow or knee of the patient to vibrate. While striking care must be taken that the strike is made at the junction of the upper 1/3 and lower 2/3 of the fork. This is the maximum vibratory area of the tuning fork. It should not be struck against metallic object because it can cause overtones. As soon as the fork starts to vibrate it is placed at the mastoid process of the patient. The patient is advised to signal when he stops hearing the sound. As soon as the patient signals that he is unable to hear the fork anymore the vibrating fork is transferred immediately just close to the external auditory canal and is held in such a way that the vibratory prongs vibrate parallel to the acoustic axis. In patients with normal hearing he should be able to hear the fork as soon as it is transferred to the front of the ear. This result is known as Positive Rinne test. (Air conduction is better than bone conduction). In case of conductive deafness the patient will not be able to hear the fork as soon as it is transferred to the front of the ear (Bone conduction is better than air conduction). This is known as negative Rinne. It occurs in conductive deafness. This test is performed in both the ears.

If the patient is suffering from profound unilateral deafness then the sound will still be heard through the opposite ear this condition leads to a false positive Rinne.

Use of Rinne test in quantifying conductive deafness:

Conductive deafness of more than 25 dB is indicated by negative Rinne with 512 Hz fork, while it is positive for 1024 Hz. If Rinne is negative for 256, 512 and 1024 Hz then conductive deafness should be greater than 40dB.

Weber test:

Weber's test:

Is a tuning fork test (quick) used to assess hearing levels in an individual. This can easily

detect unilateral conductive and unilateral sensorineural hearing loss. This test is named after Ernst Heinrich Weber (1795 – 1878).

Procedure:

Tuning forks used - 256 Hz / 512 Hz

Commonly used frequency is 512 Hz.

A vibrating fork is placed over the forehead / vertex / chin of the patient. The patient should be instructed to indicate which ear hears the sound better. In normal ear and in bilateral equally deaf ears the sound will be heard in the mid line. This test is very sensitive in identifying unilateral deafness. It can pick out even a 5 dB difference between the ears.

Theory:

A patient with a unilateral (one-sided) conductive hearing loss would hear the tuning fork loudest in the affected ear. This is because the conduction problem masks the ambient noise of the room, whilst the well-functioning inner ear picks the sound up via the bones of the skull causing it to be perceived as a louder sound than in the unaffected ear.

Inadequacies:

This test is most useful in individuals with hearing that is different between the two ears. It cannot confirm normal hearing because it does not measure sound sensitivity in a quantitative manner. Hearing defects affecting both ears equally, as in Presbycusis will produce an apparently normal test result.

Absolute Bone conduction test:

This test is performed to identify sensorineural hearing loss. In this test the hearing level of the patient is compared to that of the examiner. The examiner's hearing is assumed to be normal. In this test the vibrating fork is placed over the mastoid process of the patient after occluding the external auditory canal. As soon as the patient indicates that he is unable to hear the sound anymore, the fork is transferred to the mastoid process of the examiner after occluding the external canal. In cases of normal hearing the examiner must not be able to hear the fork, but in cases of sensorineural hearing loss the examiner will be able to hear the sound, then the test is interpreted as ABC reduced. It is not reduced in cases with normal hearing.

Bing test:

This is actually a modification of weber's test. The vibrating fork is placed over the mastoid process and when it ceases to be heard the examiner's finger is used to occlude the external auditory canal. In normal individuals the sound will be heard again. This is because by occluding the external auditory canal the examiner is preventing sound from escaping via the external canal. The external auditory canal acts as a resonating chamber. If the vibrating fork is not heard again after the external canal is occluded then it is construed that the middle ear conduction is the cause for deafness. In patients with pronounced deafness if the vibrating fork is heard after occlusion of external canal then deafness is construed to be due to labyrinthine causes.

Politzer test:

In this test the vibrating fork is held in front of open mouth and the patient is asked to swallow. If the Eustachian tubes are patent then sound will be intensified during swallowing. If only one tube is patent then sound will be accentuated only in that ear. Sometimes normal persons too may not hear the vibrating fork.

Bing Entotic test:

Hypothetically this test is supposed to differentiate between deafness due to ankylosis of foot plate of stapes from that of conditions interfering with mobility of other ossicles. This test is actually of historic value only. Eustachian catheter is passed and to one of its ends is attached a speaking tube. If the patient is able to hear the fork better via this tube than that from the external auditory canal then middle ear ossicles other than foot plate of stapes is supposed to be at fault.

Stenger's test:

This test is performed to identify feigned hearing loss and malingering. This test is based on the auditory phenomenon known as "Stenger's principle". This principle states that when two similar sounds are presented to both ears only the louder of the two would be heard. Patients usually are not aware of this phenomenon. When two similar tuning forks of same frequencies are made to vibrate and held simultaneously in the acoustic axis of both ears only the louder fork will be heard. Loudness of vibrating fork can be adjusted by adjusting the distance of the fork from the external canal. Usually the vibrating fork is held closer to the allegedly deaf ear of the patient. The patient will not acknowledge hearing in that ear. According to Stenger's principle he should be able to hear the louder fork. If the hearing loss in worse ear is genuine, patient will respond to the signal presented to the better ear.

This is known as negative Stenger's test. Feigning patient will not acknowledge hearing when louder sound is presented to the worse ear. This is known as positive Stenger's test.

Gelle test:

In this test, the air pressure in the external canal is varied using a Siegle's speculum. The vibrating fork is held in contact with the mastoid process. In normal individuals and in those with sensorineural hearing loss, increased pressure in the external meatus causes a decrease in the loudness of the bone conducted sound. In stapes fixation no alteration in the hearing threshold is evident.

Chimani-Moos test:

This is actually a modification of Weber test. When the vibrating fork is placed on the vertex, the patient indicates that he hears it in the good ear and not in the deaf ear. The meatus of the good ear is then occluded. A genuine deaf patient will still be able to lateralize the sound to the good ear, whereas a malingerer will deny hearing the sound at all.

Ventilation tubes

Introduction:

Ventilation tubes (Grommets) are prosthetic tubes inserted into the middle ear cavity via a perforation in the ear drum. These tubes serve to ventilate and drain the middle ear cavity.

Ventilation tubes can be of different materials like teflon, silicone, titanium, gold etc. These materials may be coated with silver oxide.

Features of ventilation tube:

1. It should not be irritating to the middle ear
2. It should not elicit foreign body reaction
3. It should be sterilizable
4. It should not cause allergic reaction in the individual

Ventilation tubes are of two types:

1. Grommet (dumbbell shaped): Used for short term purposes. It gets extruded within 6 months.
2. T tube (T shaped): Used for long term purposes. This tube stays for at least 1 - 2 years.

The general rule of thumb is that the larger and stiffer the flange that goes into the middle ear, the longer it stays in situ. The longer it stays, the longer gets its potential benefits. T tube is one such tube that stays for a longer period of time without extrusion. The longer the tube stays in position, greater are the chances of complications. These complications include infection, granulation tissue formation, permanent perforation of ear drum, thinning

of ear drum with possibility of retraction pockets.

Indications for ventilation tube insertion:

1. Secretory otitis media
2. Adhesive otitis media

Site of insertion: It is ideally inserted in the antero - inferior quadrant of the ear drum through a radial incision. This site is chosen due to the proximity of the Eustachian tube, and to maximize the duration of tube function.

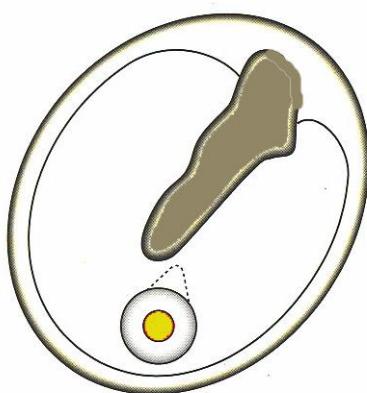
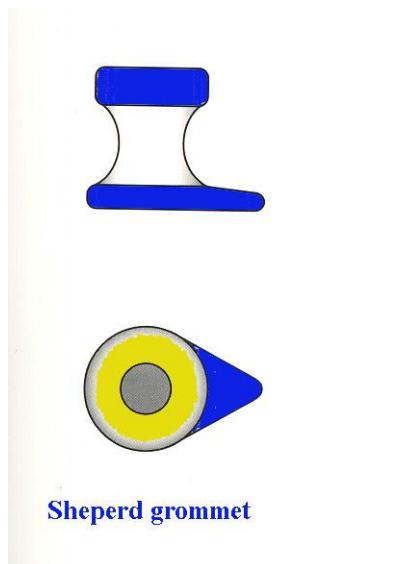


Figure showing grommet in situ

Otitis media with effusion

Synonyms: Secretory otitis media, glue ear, serous otitis media, non-purulent otitis media.

Definition: Otitis media with effusion is defined as chronic accumulation of mucus within the middle ear, and rarely this could involve the mastoid air cell system. This accumulation causes conductive hearing loss.

Histology and histopathology of Eustachian tube: The pseudostratified ciliated columnar epithelium of respiratory tract extends up the Eustachian tube as far as the anterior part of the middle ear cavity. These cells are capable of producing mucus. There are also goblet cells seen in their midst. These cells are also capable of secreting mucus material. Otitis media with effusion is caused by inflammation of this epithelium in the Eustachian tube and hypotympanum. In established cases of glue ear, the cuboidal epithelium of middle ear and mastoid air cells gets replaced by thickened pseudostratified columnar epithelium. The cilia of these cells have also been found to be ineffective in propelling the secretions into the nasopharynx. The sub mucosa is found to be oedematous, inflamed with dilated blood vessels with increased number of macrophages and plasma cells.

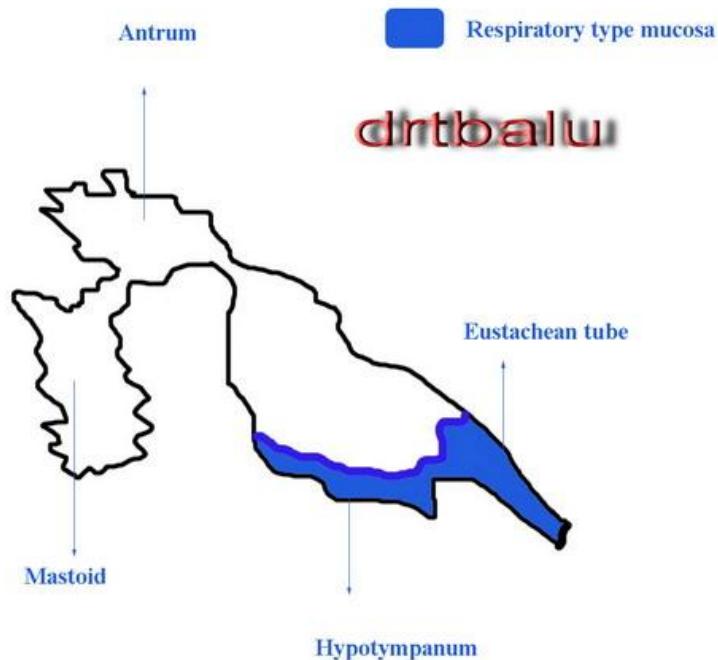


Figure showing areas lined by respiratory type of epithelium

Etiology:

1. In many children otitis media with effusion is preceded by an episode of acute otitis media. This is common in children who are more prone for upper respiratory infections. Common being viral infections which damages the Eustachian tube epithelium.
2. Craniofacial abnormalities: Children with cleft palate have deficient palatal muscles causing a poor Eustachian tube function leading on to Otitis media with effusion. This occurs despite a successful surgical repair of the cleft palate. Children with Down's syndrome are also more prone for OME.
3. Allergy: Previously nasal allergy has been postulated as an important factor in the development of Otitis media with effusion. Studies have been unequivocal.

Note: Children with bifid uvula do not appear to have higher incidence of OME

4. Gastro oesophageal reflux: GERDS has been commonly demonstrated radiologically in children with OME. Furthermore biochemical analysis of middle ear fluid has demonstrated significant amounts of pepsin (in 80% of cases).

5. Parental smoking has been attributed as an important predisposing factor for the development of OME.

Age of occurrence: OME shows classically a bimodal distribution. The first peak occurs around 2 years of age, and the second peak occurs at about 5 years of age. This distribution occurs roughly around the ages when the child goes to preschool and primary school.

Seasonal association: OME commonly occurs during winter season, when there is more likelihood of upper respiratory infections, and also because of the possibility of closer contact with affected children. This is seen in temperate zones. In non-temperate zones it is commonly seen during rainy season.

Clinical features: A high index of suspicion is necessary to identify this condition. Every child with upper respiratory infection must be otoscopically examined.

Otoscopic findings: The tympanic membrane may be bulging, or retracted with a distorted cone of light. The ear drum may appear yellow, blue or simply clear white. Pneumatic otoscopy will reveal a ear drum which has a restricted mobility.



Otoscopic picture of ear drum with middle ear effusion

Investigations:

Pure tone audiometry: Demonstrates mild to moderate conductive deafness.

Tympanograms (Type B) is commonly associated with OME. Type A is infrequently associated while Type C falls somewhere in between. Tympanometry can be used as a screening test to identify patients with OME.

Free field audiometry: Demonstrates deafness.

Management:

1. Antibiotics: Amoxycillin is the drug of choice followed by cephalosporins.

2. Nasal decongestants like oxymetazoline / xylometazoline may help in some cases.

3. Topical nasal steroids can be used in resistant cases.

4. Auto inflation of Eustachian tube by performing Valsalva manoeuvre. Balloon blowing may also help.

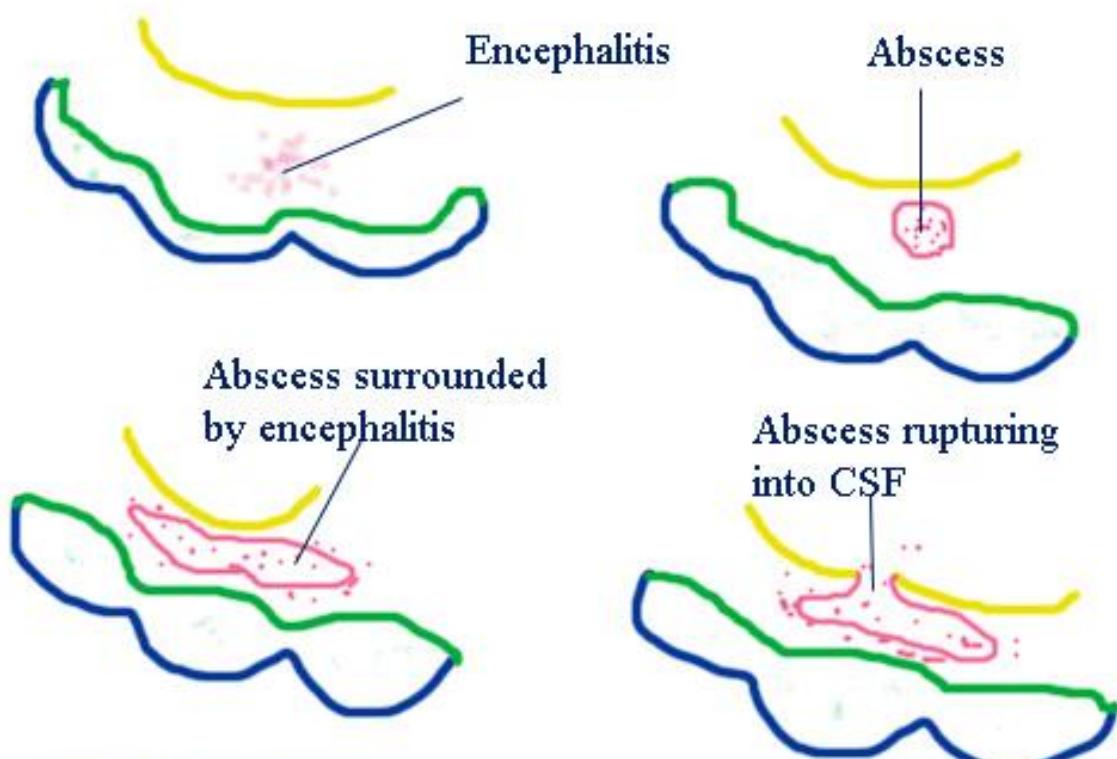
Surgical management:

1. Adenotonsillectomy

2. Myringotomy and insertion of ventilation tubes

Otogenic brain abscess ¹

Otogenic brain abscess always develop in the temporal lobe or the cerebellum of the same side of the infected ear. Temporal lobe abscess is twice as common as cerebellar abscess. In children nearly 25% of brain abscesses ² are otogenic in nature, whereas in adults who are more prone to chronic ear infections the percentage rises to 50%. The routes of spread of infection has already been discussed above, the commonest being the direct extension through the eroded tegmen plate. Although dura is highly resistant to infection, local pachymeningitis may be followed by thrombophlebitis penetrating the cerebral cortex, sometimes the infection could extent via the Virchow - Robin spaces in to the cerebral white matter. Cerebellar abscess is usually preceded by thrombosis of lateral sinus. Abscess in the cerebellum may involve the lateral lobe of the cerebellum, and it may be adherent to the lateral sinus or to a patch of dura underneath the Trautmann's triangle.



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Diagram showing evolution of brain abscess

Stages of formation of brain abscess:

Stage of cerebral oedema: This is in fact the first stage of brain abscess formation. It starts with an area of cerebral oedema and encephalitis. This oedema increases in size with spreading encephalitis.

Walling off of infection by formation of capsule stage: Brain attempts to wall off the infected area with the formation of fibrous capsule. This formation of fibrous tissue is dependent on microglial and blood vessel mesodermal response to the inflammatory process. This stage is highly variable. Normally it takes 2 to 3 weeks for this process to be completed.

Liquefaction necrosis: Infected brain within the capsule undergoes liquefactive necrosis with eventual formation of pus. Accumulation of pus cause enlargement of the abscess.

Stage of rupture: Enlargement of the abscess eventually leads to rupture of the capsule containing the abscess and this material finds its way into the cerebrospinal fluid as shown in the above diagram.

Cerebellar abscess which occupy the posterior fossa cause raised intra cranial tension earlier than those above the tentorium. This rapidly raising intra cranial pressure cause coning or impaction of the flocculus or brain stem into the foramen magnum. Coning produces impending death. If the walling off process (development of capsule) is slow, softening of brain around the developing abscess may allow spread of infection into relatively avascular white matter, leading to the formation of secondary abscesses separate from the original or connected to the original by a common stalk. This is how multilocular abscesses are formed. Eventually the abscess may rupture into the ventricular system or subarachnoid space, causing meningitis and death.

The mortality rate of brain abscess is around 40%, early diagnosis after the advent of CT scan has improved the prognosis of this disease considerably..

The bacteriological flora is usually a mixture of aerobes and obligate anaerobes. Anaerobic streptococci are the commonest organisms involved. Pyogenic staphylococci is common in children. Gram negative organism like proteus, E coli and Pseudomonas have also been isolated.

Clinical features:

The earliest stage where the brain tissue is invaded (stage of encephalitis) is marked by the presence of headache, fever, malaise and vomiting. Drowsiness eventually follows. These early features may be masked by the complications such as meningitis or lateral sinus thrombosis. If this stage progresses rapidly to generalised encephalitis before it could be contained by the formation of the capsule, drowsiness may progress to stupor and coma followed by death. Usually the period of local encephalitis is followed by a latent period

during which the pus becomes contained within the developing fibrous capsule. During this latent phase the patient may be asymptomatic.

During the next state (stage of expansion) the enlarging abscess first cause clinical features due to the alteration of CSF dynamics, and site specific features may also be seen due to focal neurological impairment. The pulse rate slows with rising intracranial pressure, the temperature may fall to subnormal levels. Drowsiness may alternate with periods of irritability. Papilledema is also found due to elevated CSF pressure.

Clinical features also vary according to the site of involvement. This accounts for the differences that are seen between the cerebral and cerebellar abscess.

Cerebral (Temporo sphenoidal abscess):

A cerebral abscess in the dominant hemisphere often causes nominal aphasia, where in the patient has difficulty in naming the objects which are in day to day use. He clearly knows the function of these objects. Visual field defects arise from the involvement of optic radiations. Commonly there is quadrantic homonymous hemianopia, affecting the upper part of the temporal visual fields; more rarely it may also involve the lower quadrants. The visual field losses are on the side opposite to that of the lesion. This can be assessed by confrontation method. Upward deviation affects facial movements on the opposite side, and then progressively paralysis of the upper and lower limbs. If the expansion occurs in inward direction then paralysis first affects the leg, then arm and finally the face.

Cerebellar abscess:

The focal features associated with cerebellar abscess are weakness and muscle incoordination on the same side of the lesion. Ataxia causes the patient to fall towards the side of the lesion. Patient may also manifest intention tremors which may become manifest by the finger nose test. This test is performed by asking the patient to touch the tip of the nose with the index finger first with the eyes open and then with the eyes closed. The patient may often overshoot the mark when attempted with the eyes closed in case of cerebellar abscess. The patient may also have spontaneous nystagmus. Dysdiadokinesis is also positive in these patients.

Investigations:

CT scan and MRI scans are the present modes of investigation. Scan is ideally performed using contrast media. These scans not only reveal the position and size of the abscess, the presence of localised encephalitis can be distinguished from that of an encapsulated abscess. Associated conditions such as subdural abscess, and lateral sinus thrombosis can also be seen.

Management:

Surgical drainage of the abscess, followed by mastoidectomy to clear the ear disorder is the preferred management modality.

1.

Otogenic brain abscesses - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 27];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/otogenic-brain-abscesses>

2.

Complications of otitis media by drtbalu [Internet]. [cited 2011 Dec 28];Available from: http://www.drtbalu.co.in/compli_om.html

Keratosis obturans ¹

Keratosis obturans: is accumulation of desquamated keratin in the external auditory meatus. This should be differentiated from primary auditory canal cholesteatoma which is characterized by invasion of squamous tissue from the external ear canal into a localized area of bone erosion.

Pathology: The keratin plug seen in keratosis obturans appears like a geometrically patterned keratin plug within the lumen of expanded ear canal. These keratin squames are shed from the complete circumference of the deep ear canal forming a lamina. It appears like onion skin.

Etiology: Keratosis obturans is postulated to occur due to abnormal epithelial migration of ear canal skin. The movement of the surface epithelium appears to be reversed in these patients. (The surface epithelium over pars flaccida migrates downwards to the pars tensa and then moves inferiorly across the drum).

Keratosis tympanicum: Is also caused by abnormal migration of squamous epithelium lining the deep portion of the external auditory canal. This condition is also associated with unilateral tinnitus.

Types of keratosis obturans:

- a. **Inflammatory type:** This is caused due to acute inflammation involving the external ear canal. Viral infections commonly cause this problem. The inflammatory reaction involving the ear canal temporarily alters epithelial migration. This condition can only be cured by removal
- b. **Silent type:** In this type there are no predisposing acute infections involved. This condition is postulated to be caused by abnormal separation keratin that persists even after the removal, and will need repeated removals.
- c. **Primary auditory canal cholesteatoma:** Etiology is uncertain. It is commonly thought to be caused by trauma to the bone covering the external canal. This could also be caused by surgical trauma as in patients who have undergone stapedectomy. The piece of exposed bone in the external canal becomes infected and sequesters. The lining epithelium migrates into this area causing the formation of cholesteatoma. This condition is characterized by ear pain which is dull and aching in nature. It is not associated with hearing impairment.

Keratosis obturans commonly occur in young patients.

Clinical features:

1. Severe ear pain

2. Mild / moderate conductive hearing loss
3. Associated bronchitis / sinusitis - common

On examination:

The ear canal appears to be widened, making the ear drum stand out. CT scan of temporal bones may reveal canal erosion and widening.

After surgical removal under general anesthesia the specimen must be sent for pathological evaluation to rule out malignancy.

Management:

1. Surgical removal under G.A.
2. Canal plasty is helpful in recurrent cases
3. Mastoidectomy should be performed in cases with primary cholesteatoma of external canal.

1.

Keratosis obturans - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 28]; Available from: <https://sites.google.com/site/drtbalusotolaryngology/otology/keratosis-obturans>

Myringitis granulosa ¹

Definition: Granular myringitis (Myringitis granulosa) is a specific form of otitis externa. It is characterized by the presence of granulation tissue on the lateral aspect of the ear drum sometimes with involvement of external auditory canal.

Synonyms:

1. Granular myringitis
2. Granulating myringitis
3. Granulomatous otitis externa
4. Chronic myringitis
5. Acute granulomatous myringitis

Histopathology: Specimen for HPE is easy to obtain.

Features:

1. Oedematous granulation tissue with capillaries and diffuse infiltration of chronic inflammatory cells.
2. There is no lining epithelium over these granulation tissue

Etiology:

1. High ambient temperature
2. Swimming
3. Lack of personal hygiene
4. Exposure to local irritants
5. Foreign bodies
6. Bacterial and fungal infections
7. Occasionally may complicate grafted ear drum

Symptoms:

1. Foul smelling discharge from the involved ear
2. There is very little or negligible pain
3. There may be fullness / irritation in the affected ear
4. Hearing is nearly normal
5. Some patients may be totally asymptomatic

Signs:

The ear drum is covered with purulent secretions, which on removal reveals the underlying granulation tissue. Granulations may be localized or diffuse. The localized form of granulations are the most common, here small areas of the drum are affected, with formation of one or more polyps. These granulations are common over the postero superior

margin of the ear drum. These granulations may also affect the adjacent wall. Despite all these signs the ear drum is intact.

All these patients must undergo pure tone audiometric evaluation to rule out middle ear pathology.

Pathophysiology:

Myringitis can be a self-maintained primary disorder of ear drum (primary myringitis) or due to inflammatory conditions involving adjacent areas like external canal and middle ear cavity (secondary myringitis).

Causes of primary myringitis:

1. Direct trauma to ear drum due to foreign body
2. Unsuccessful attempts at removal of foreign body
3. Bacterial infections (streptococcal pneumonia)
4. Influenza infections
5. Herpes infections
6. Eczematous myringitis due to dermal eczema of overlying ear drum's epidermis

Causes of secondary myringitis:

1. Acute myringitis due to acute otitis media
2. Acute myringitis due to otitis externa

Management:

Localized form: Meticulous microscopic debridement will help.

Topical administration of steroid / antibiotic ear drops will help.

These patients should be treated over a prolonged period of time.

Application of caustic agents like formalin / Tricholoroacetic acid can be tried.

In refractory cases surgical removal of granulations can be attempted.



Figure showing myringitis granulosa

1.

Balasubramanian T drtbalu. Myringitis granulosa [Internet]. drtbalu's otolaryngology online. 2011 Dec 28; Available from:

<https://sites.google.com/site/drtbalusotolaryngology/otology/myringitis-granulosa>

Perichondritis Pinna ¹

Definition: This term refers to infection / inflammation involving the perichondrium of the external ear, which includes the auricle and external auditory canal. It is commonly used to describe a series of conditions of external ear ranging from erysipelas (infections of skin), cellulitis (infections of soft tissue), and true Perichondritis to chondritis (infections involving the cartilage).

Classification:

- Erysipelas of external ear
- Cellulitis of external ear
- Perichondritis
- Chondritis

Etiology:

- It commonly occurs due to trauma. Trauma includes laceration of auricle, surgery to external ear, frost bite, burns, chemical injury, high piercing of ear lobe to insert ear rings, infection of hematoma of pinna.
- Superficial infections of skin lining (erysipelas)
- Infections involving subcutaneous tissue (cellulitis)
- Infections involving the perichondrium (Perichondritis)
- Infections of cartilage (cartilage)

Microbiology:

Organisms commonly isolated include *Pseudomonas aeruginosa* and *staphylococcus aureus*.

Clinical features:

Dull aching pain involving the cartilaginous portion of pinna. The ear lobule which is devoid of cartilage is spared. Classic signs of inflammation are clearly demonstrable. The pain is more severe in perichondritis when compared to erysipelas or cellulitis.

This condition should be differentiated from relapsing polychondritis. Relapsing polychondritis is associated with systemic effects like fever.

Perichondritis when left untreated causes necrosis of the underlying cartilage causing deformities of pinna (cauliflower ear).

Management:

Prevention: Perichondritis can be prevented by placing the ear prick sites well away from the cartilage of the pinna (over the ear lobule).

Hematomas of auricle should be drained immediately following aseptic precautions.

Burn injuries of pinna should be carefully managed.

Mild forms of perichondritis can be managed by a course of broad spectrum antibiotics, administered preferably in high doses. Subperichondrial abscess should be drained immediately.

In resistant and recurrent cases Button surgery can be performed.

Continuous drainage of subperichondrial abscess can be resorted to in resistant cases by placing polythene tubes over the site of incision. Topical Antibiotics can also be administered through these tubes. Topical antibiotics can be of use because the cartilage is devoid of blood supply and systemic antibiotics may not reach the cartilage in adequate concentrations.

Highly resistant cases may be managed by low dose radiation which could kill the micro-organisms. Three or four sittings of 0.8 Gy radiation may be used over a period of two days.



Image showing perichondritis

Citations:

1.

Balasubramanian T drtbalu. Perichondritis pinna [Internet]. Perichondritis pinna. 2011 Dec 28;Available from:
<https://sites.google.com/site/drtbalusotolaryngology/otology/perichondritis-pinna>

Unilateral otalgia ¹

Unilateral pain in the ear can be classified into:

- i. External ear causes
- ii. Middle ear causes
- iii. Pain referred from adjacent areas due to segmental innervation.

External ear causes: Otitis externa is the most common cause of unilateral pain in the ear. This is due to infection of the external auditory canal. This condition is really a very painful one.

Otitis external can be classified into:

1. Acute diffuse otitis externa (commonly caused by bacteria)
2. Acute localised otitis externa (commonly furuncle)
3. Chronic otitis externa
4. Eczematous otitis externa
5. Fungal otitis externa
6. Malignant otitis externa

Predisposing factors for otitis externa:

Under normal conditions the skin lining the external auditory canal is well protected by its self-cleansing mechanism. In diseased conditions several factors may come into play in the pathogenesis of otitis externa.

1. Absence of cerumen: The cerumen plays an important role in the protection of the external canal. It protects the external canal from moisture. It also has anti-bacterial properties which helps in the protection of the external canal. The cerumen also lowers the pH of the external canal making it difficult for the bacterial pathogens to colonize.
2. Removal of cerumen by ear buds: is one of the common causes of otitis externa. The act of removal traumatises the skin lining of the external canal making it vulnerable to infections.
3. Frequent exposure to water: external canal when constantly bathed in water loses its ability to protect itself. The presence of water macerates the skin lining of the external canal and also increases the pH of the external canal making it more favourable for bacterial colonisation. This condition is common in swimmers.

Acute diffuse otitis externa:

This is also known as the swimmers ear. This is an inflammatory condition involving the external canal in a diffuse manner. This condition is common in swimmers because of the propensity for the external canal to be exposed to water for long durations. This exposure leads to maceration of the external canal skin, and also lowers the pH of the external canal providing an environment favourable to infections.

Main symptoms:

1. Itching in the external canal
2. Tenderness on palpation
3. Aural fullness rarely occur due to the reduction in size of the external canal lumen due to oedema
4. Rarely stenosis of the external canal may occur causing accumulation of debris and secretions

Common signs:

1. Erythema of the external canal
2. Oedema of external canal
3. Secretions from the external canal (weeping canal)
4. Pain on mastication
5. Pulling of helix in a postero superior direction cause pain
6. In advanced cases fever and lymphadenopathy may occur (pre and post auricular nodes may be involved)

Stages of acute diffuse otitis externa: (Senturia)

Preinflammatory stage: is characterised by intense itching, oedema and sensation of fullness in the ear.

Inflammatory stage: may be divided into mild, moderate and severe.

Mild acute inflammatory stage: here the cardinal features are increased itching, pain, mild erythema and oedema of the external canal skin. At later stages exfoliation of skin with minimal amount of cloudy secretions may be seen in the external canal.

Moderate acute inflammatory stage: in this stage the itching and tenderness of the external canal intensifies. The external canal is narrowed due to oedema and accumulation of epithelial debris.

Severe acute inflammatory type: In this stage pain becomes intolerable to such an extent the patient may refuse to eat, the lumen of the external canal becomes totally obliterated due to oedema and accumulated epithelial debris. Otorrhoea may become purulent. In addition regional nodes may also be involved. Infections from the external canal may involve the parotid gland via the fissure's of santorini.

Common organisms involved: Psuedomonas aeruginosa and staphylococcus aureus are commonly cultured from the external canal of these patients. The normal commensals like staphylococcus epidermidis and corynebacteria are conspicuously absent.

Management:

The aim is twofold:

1. Resolving the infection
2. Promoting the external canal skin's recovery to its original state.

Firstly the canal is cleaned atraumatically by gentle suctioning and debridement under microscope. Topical hydrogen peroxide solution instilled will help the process of debridement.

A cotton wick dipped in I.G. paint can be inserted in to the external canal and allowed to stay for a day. This will reduce the external canal skin oedema and will increase the size of the meatus. Ear drops containing a mixture of neomycin and 1% hydrocortisone may be instilled as ear drops at least three times a day. In addition to the antibiotic and anti-inflammatory effects this drug reduces the pH of the external canal making it more resistant to the organisms.

In severe cases oral antibiotics and anti-inflammatory drugs can be resorted to. Quinolones are commonly used oral antibiotic.

Acute localized otitis externa:

This condition is otherwise known as furunculosis or circumscribed otitis externa. This is a localised infection usually found to involve the lateral 1/3 of the external canal. It also has a propensity to involve the posterior superior aspect of the external canal. This is caused due to obstruction of the apopilosebaceous units found extensively in this area.

Trauma to skin in this area followed by infection is commonly attributed cause. The organism responsible is commonly staph aureus.

Symptoms:

1. Localised pain
2. Localised itching
3. Purulent discharge if the abscess ruptures
4. If oedema or abscess occludes the external canal hearing loss can occur.

Signs:

1. Erythema of the skin
2. Localised abscess formation

Management:

If the abscess is pointing it can be treated by incision and drainage. Oral antibiotics should be used. The preferred drug of choice is penicillin of first generation cephalosporins. Anti-inflammatory drugs can be used to reduce inflammation and pain.

These patients must be advised to cut their nails short and to keep their hands clean, since this is the commonest route of infection.

Chronic otitis externa:

This is a chronic infection / inflammation involving the skin lining of the external canal. There is thickening of the skin lining of the external canal due to persistent low grade infection / inflammation.

Symptoms:

1. Unrelenting pruritus
2. Mild pain
3. Presence of dry skin in the external canal

Signs:

1. Asteatosis (lack of cerumen)
2. Hypertrophic external canal skin
3. Presence of dry flaky skin in the external canal

4. Mild tenderness on ear manipulation

5. Rarely muco purulent Otorrhoea

Cultures from the external canal of these patients are highly unreliable because they would have been using various antibiotic drops to surmount the problem.

Management:

Involves extensive use of acetic acid ear drops. This helps to reduce the pH of the skin lining the external canal making it more resistant to bacterial infections. In intractable cases steroid drops can be tried. Antibiotic drops may not be useful in these patients.

Surgery is indicated in extreme cases. A canalplasty is performed to widen the external canal. The involved skin may be removed to be replaced by a split thickness graft.

Eczematous otitis externa:

This condition includes various dermatologic conditions involving the skin of the external canal. It may range from atopic dermatitis, contact dermatitis, seborrhoea dermatitis, neuro dermatitis, infantile eczema etc.

This condition is characterised by intense itching, in fact this could be the only complaint of the patient. On examination, erythema of the external canal skin may be seen. There may also be associated scaling and oozing from the canal skin.

Success lies in the management of the underlying dermatologic condition

Fungal otitis externa:

This is the commonest type of otitis externa in tropical countries. This condition is associated with increased ear canal moisture, or following treatment of otitis external by prolonged use of topical antibiotics. The protective cerumen layer is absent in these patients. This condition is more common in diabetics.

Symptoms:

1. Intense itching

2. Pain when otitis externa is coexistent

3. Blocking sensation due to the presence of fungal balls

Signs:

1. Inflamed external canal skin

2. External canal tenderness

3. Fungal debris (black in case of aspergillus and white in the case of candida). Invariably the infection is mixed type.

Management:

The condition is managed by careful aural toileting to remove the fungal balls. The best way to remove fungus from the ear canal is by aural syringing. Antifungal ear drops of clotrimazole can be administered. If secondary infections are present oral antibiotics and anti-inflammatory drugs may be resorted to.

Malignant otitis externa:

This rare but sinister form of otitis externa is known to affect elderly diabetics. This condition is caused by pseudomonas infection of the external ear. These patients have a unique nocturnal deep boring type of pain.

The patient gives history of trivial trauma to the external canal. Granulations can be seen at the junction of bony cartilaginous portion of the external canal.

This condition can cause complications like facial nerve involvement, and spread to the intracranial structures.

Middle ear causes of otalgia:

Acute otitis media is one of the common middle ear causes of otalgia. This condition is common in children. It is caused by Eustachian tube block causing pent up secretions to accumulate in the middle ear cavity. Pain gets relieved when the ear drum perforates and starts to drain the middle ear cavity. Children are commonly affected because of their short, wide and straight Eustachian tube.

Otitic barotrauma: This is caused due to sudden changes in altitudes as in deep sea diving / flying unpressurized airplanes.

Referred otalgia:

Pain to the ear can be referred from disorders affecting other portions of head and neck. These include:

1. Temporomandibular joint dysfunction
2. Dental pain
3. Quinsy
4. Tonsillitis
5. Post tonsillectomy pain always radiates to the ipsilateral ear.

Causes for referred otalgia should be diligently searched for in a patient with ear pain, with clinically normal ear.

Citations:

1.
Otalgia unilateral - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 28];Available from:
<http://sites.google.com/site/drtbalusotolaryngology/otology/otalgia-unilateral>

Role of Maskers in tinnitus management ¹

Role of tinnitus maskers in the management of tinnitus:

Tinnitus maskers play a vital role in the management of tinnitus. Use of tinnitus maskers reduces the incidence of tinnitus in these patients.

Types of tinnitus maskers:

There are three different masking devices used in the management of tinnitus. The exact type used depends on the degree of patient's hearing loss, and the nature of tinnitus.

Type I: Conventional hearing aids. These conventional hearing aids produce masking of tinnitus by amplifying ambient noise which in turn covers up or masks the tinnitus. These hearing aids are useful in patients who have tinnitus associated with hearing loss.

Type II: These are classic tinnitus maskers that generate noise bands which can be adjusted by the user. The user can choose the noise band and the optimal volume that masks the tinnitus

Type III: Tinnitus instruments. These are a combination of both a high frequency hearing aid and a tinnitus masker within the same apparatus. Both these components have separate volume controls that can be used to adjust them individually.

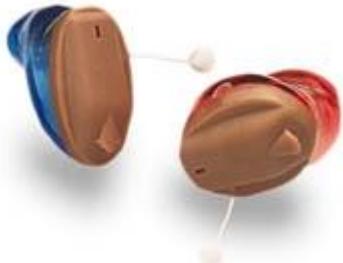


Figure showing tinnitus masker

Indications of various tinnitus maskers:

1. Patients with normal hearing commonly have high frequency hearing loss and hence should use tinnitus maskers that generate masking sounds with substantial high frequency content (6kHz and above).
2. Patients with high frequency hearing loss associated with high pitched tinnitus should use tinnitus instruments (type III).
3. Patients with low frequency hearing loss with associated low pitch tinnitus would benefit from using well fitted hearing aids. These hearing aids are sufficient to produce masking effect on the tinnitus.

It is always important to adjust the tinnitus masker so as to generate the lowest level of masking sound that is capable of masking the tinnitus.

If tinnitus is troublesome during sleep then bedside maskers can be used. Masking pillows are also available.

Tinnitus masking by using bone conduction of ultrasound: One recent advancement in tinnitus masking is the development of new wearable high frequency bone conduction maskers. These maskers can be applied anywhere over the skull without the need to occlude the external auditory meatus.

Citation:

1.
Role of maskers in tinnitus management - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 28];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/role-of-maskers-in-tinnitus-management>

Siegel's speculum

A siegel's pneumatic speculum has an eye piece which has a magnification of 2.5 times. It is a convex lens. The eye piece is connected to an aural speculum. A bulb with a rubber tube is provided to insufflate air via the aural speculum. The advantages of this aural speculum is that it provides a magnified view of the ear drum, the pressure of the external canal can be varied by pressing the bulb thereby the mobility of ear drum can be tested. Since it provides adequate suction effect, it can be used to suck out middle ear secretions in patients with CSOM. Ear drops can be applied into the middle ear by using this speculum. Ear is first filled with ear drops and a snugly fitting siegel's speculum is applied to the external canal. Pressure in the external canal is varied by pressing and releasing the rubber bulb, this displaces the ear drops into the middle ear cavity.



Figure showing siegel's speculum

Tinnitus retraining therapy

Tinnitus retraining therapy: was first introduced by Jastreboff during 1980. This therapy has been found to be highly successful. This therapy is based on strong neurophysiologic evidence that any person can habituate to acoustic or acoustic like sensations in their environment.

The major aim of this treatment protocol is to train the brain to treat tinnitus like any other routine environmental sound (like the hum of refrigerator in the kitchen which does not bother them).

Tinnitus retraining therapy has two key elements: They are

1. Direct counselling
2. Sound therapy

Direct counselling: The counselling session is very critical to the success of this programme. Some patients may actually achieve relief through counselling alone. The counselling process involves an in depth discussion with the patient regarding the physiology of hearing and tinnitus. The negative emotions associated with tinnitus could easily be removed by counselling.

Sound therapy: These patients are fitted with white noise devises at the ear level. These devises look like small hearing aids and can be comfortably worn throughout the day. The sound is initially set at a very low level so as not to interfere with normal hearing. After several weeks of use most patients do not hear the sound unless they really try to hear it. These devises help the brain to ignore the random signals of tinnitus. This process is known as auditory habituation.

Within 6 - 24 months many patients are fully benefited.

Role of alprazolam in the management of tinnitus ¹

Alprazolam: the commonly used anxiolytic drug has demonstrated beneficial effects in the management of tinnitus. This was in fact an accidental finding when a large number of patients who were suffering from anxiety and tinnitus were prescribed this drug to relieve anxiety. A majority of them reported relief from tinnitus also. At first this beneficial effect was attributed to the fact that a less anxious patient will be able to handle tinnitus better. Further studies have without doubt demonstrated the beneficial effects of the drug.

Dosage regimen: There is a high degree of variability in the patient response to tinnitus and the dose of drug administered. After a lot of trial and error the following dosage schedule has been evolved:

1. Weeks 1 and 2 - 0.5 mg alprazolam is prescribed each evening before bed time. This dose is not sufficient to relieve tinnitus, but it allows patients to adapt to the drowsiness that is associated with the drug usage. Patients should be warned about this problem of drowsiness.
2. Weeks 3 and 4 - 0.5 mg alprazolam is given in twice a day dose (morning and evening). If this dose fails to relieve tinnitus then this drug should be continued in the same dose indefinitely, under supervision of the physician.
3. Weeks 5 and 6 - the dosage administered can also be increased to thrice a day. If the patient derives benefit the patient can continue the drug indefinitely under supervision
4. If the benefit is not sufficient to the patient's liking then the drug can be tapered off and discontinued. The tapering dose is as follows:
0.5 mg aprazolam twice a day for 3 days and then once a day for 3 days. Eventually the drug is stopped on the 7th day.

Citation:

1.
Role of alprazolam in the management of tinnitus - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 28]; Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/role-of-alprazolam-in-the-management-of-tinnitus>

Pars flaccida retraction pockets

Introduction: Pars flaccida retractions have a vital role to play in the pathophysiology of cholesteatoma. Tos et al classified pars flaccida retraction in to four stages.

Stage I: Pars flaccida is dimpled and is more retracted than normal. It is not adherent to the malleus.

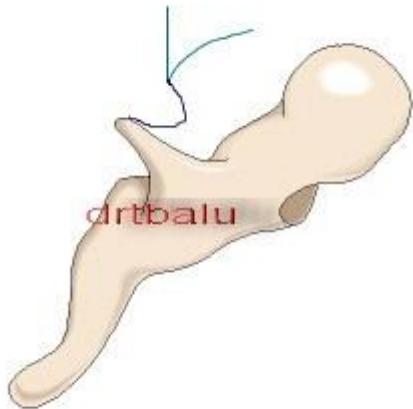


Figure showing Toss I retraction

Stage II: In this stage the retraction pocket is adherent to the handle of malleus. The full extent of the retraction pocket can be clearly seen.

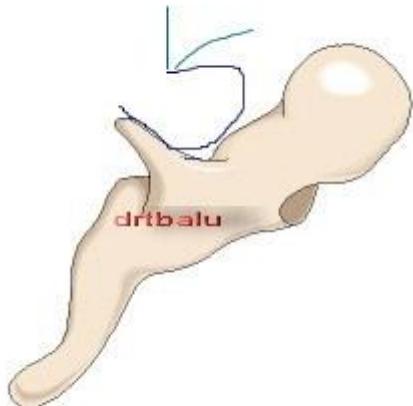


Figure showing Toss II retraction

Stage III: In this stage part of the retraction pocket may be hidden. There may also be associated erosion of the outer attic wall (scutum).



Figure showing Toss III retraction

Stage IV: In this stage there is definite erosion of the outer attic wall. The extent of the retraction pocket cannot be clearly seen as most of it are hidden from the view.

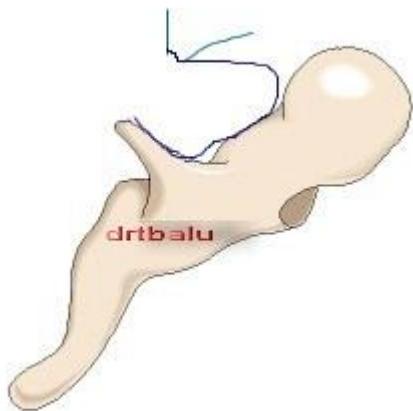


Figure showing Toss IV retraction

This classification described by Toss is fairly simple to apply, the only difficulty being the difficulty in making a distinction between stages 3 and 4. Hence for practical purposes these two stages are grouped together.

Impedance matching mechanism of middle ear cavity

Impedance matching is one of the important functions of middle ear. The middle ear transfers the incoming vibration from the comparatively large, low impedance tympanic membrane to the much smaller, high impedance oval window.

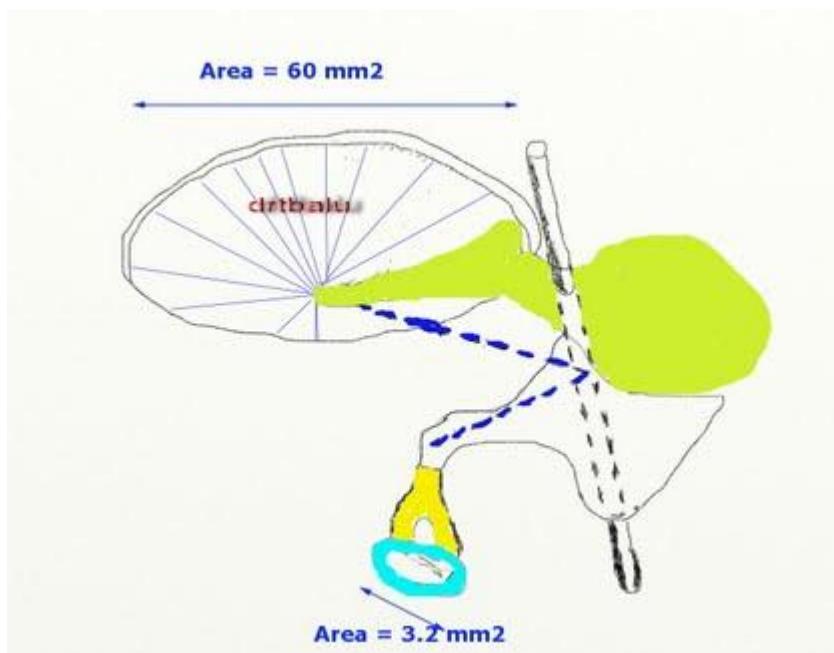
Middle ear is an efficient impedance transformer. This will convert low pressure, high displacement vibrations into high pressure of the air into, low displacement vibrations suitable for driving cochlear fluids.

The impedance of cochlear fluids is approximately equal to that of sea water (i.e. 1.5×10^6 N.sec/m³). Because of this high impedance of cochlear fluids only 0.1 % of incident energy would be transmitted.

Two processes are involved in the impedance matching mechanism of middle ear. They are:

1. The area of the tympanic membrane is larger than that of the stapes foot plate in the cochlea. The forces collected over the ear drum are concentrated over a smaller area, thus increasing the pressure over oval window. The pressure is increased by the ratio of these two areas i.e. 18.75 times.

2. The second process is the lever action of the middle ear bones. The arm of the incus is shorter than that of the malleus, and this produces a lever action that increases the force and decreases the velocity at the stapes. Since the malleus is 2.1 times longer than the incus, the lever action multiplies the force by 2.1 times.



Carhart's Notch

Carahart's notch: Is classically found in bone conduction audiograms of patients with otosclerosis. This is actually a dip centred around 2000 Hz.

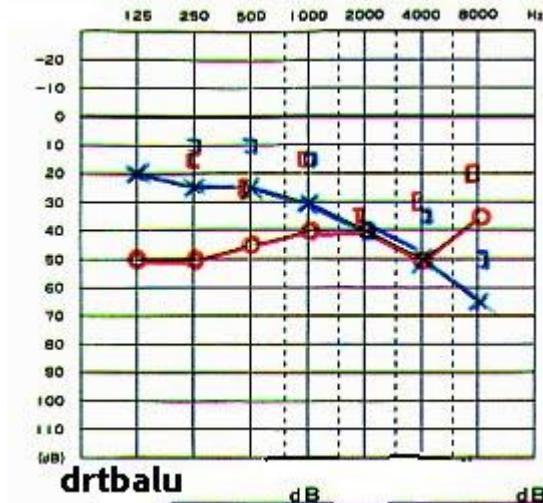


Figure showing carhart's notch

some authors consider carhart's notch to be an artefact. This notch is closely related to the carhart's effect.

Carhart's effect: was initially described following successful stapes surgery. There was an over closure of air bone gap following successful surgery. Classically this effect lead to an improvement in hearing levels particularly at 2 KHz frequency levels.

How carhart's effect is created?

When skull is vibrated by bone conduction, sound is transferred to cochlea via three routes. i.e.

1. By direct vibration of skull
2. By vibration of ossicular chain which is suspended within the skull
3. By transmission via external auditory canal (normal route)

In conductive hearing loss routes 2 and 3 are affected, but can be regained following successful stapes surgery. Hence bone conduction thresholds improve around 2 KHz frequency range.

Medical Management of Meniere's disease ¹

Medical management of Meniere's disease includes: ²

1. Dietary management
2. Physiotherapy
3. Psychological support
4. Pharmacologic intervention

Dietary management:

This includes reduction of sodium in the diet. In fact it was Frustenberg in 1934 who introduced a low salt diet for patients with Meniere's disease. Pathophysiology of Meniere's disease is enlargement of membranous labyrinth due to excess accumulation of endolymphatic fluid. Any attempt to reduce this fluid level will help in alleviate the symptoms of the patient.

Medical management is mainly used to treat patients during the acute phase of the attack. Vestibular suppressants are commonly used. Drugs used to control attacks of vertigo have varying levels of anticholinergic, antiemetic and sedative properties. Drugs used to alleviate symptoms include phenothiazines (prochlorperazine and perphenazine), antihistamines like (cinnarizine, cyclizine, dimenhydrinate, and meclizine hydrochloride), benzodiazepines like (lorazepam and diazepam).

Vestibular suppressants:

Diazepam: when used acts as vestibular depressant. It also alleviates the anxiety associated with this disorder. The beneficial effects of diazepam on the vestibular system is presumed to be due to an increase in the cerebellar GABA-ergic system. Stimulation of cerebellar GABA-ergic system mediates inhibition on the vestibular response. This drug is very useful in alleviating vertigo especially when associated with anxiety. Usual dose is 5 mg administered orally every 3 hours. The initial dose may also be administered intravenously.

Antiemetic drugs:

Drugs belonging to this group help to alleviate vomiting in Meniere's disease.

Anticholinergic drugs:

Glycopyrrolate an anticholinergic drug when combined with diazepam is helpful in controlling inner ear symptoms of nausea and vomiting. In adults it is administered in doses of 1-2 mg. It may also be administered as intramuscular injection (0.1 - 0.2 mg) every 4 hours. A side effect (reversible) of this drug includes dry mouth, distortion of visual acuity, exacerbation of symptoms in patients with prostatic hypertrophy. This drug is contraindicated in patients with glaucoma and prostatic hypertrophy.

Antidopaminergic drugs:

Droperidol: This is an antidopaminergic drug used to alleviate the symptoms of Meniere's disease. This drug is administered in doses of 2.5 - 10 mg orally in adults. If administered intravenously it is given as 5 mg bolus. This drug has fewer incidence of side effects like extrapyramidal symptoms / sedation / hypotension.

Prochlorperazine: This drug belongs to phenothiazine group. It is used as an antiemetic and a potentiator of analgesic and hypnotic drugs. Usual recommended dose is 10 mg given orally or intramuscularly every 4 - 6 hours in adults. This drug has excellent antiemetic effect.

Antihistamines:

Dimenhydrinate: is useful in preventing and treating vertigo associated with Meniere's disease. It is also very effective in controlling nausea and vomiting. Only side effect of this drug is its propensity to cause drowsiness. It is administered as 50 - 100 mg doses thrice a day. This drug can also be administered intramuscularly / intravenously.

Diphenhydramine: This drug is not useful in treating acute vertigo. It may be useful in prevention of vertigo. The usual duration of action is 4-6 hours. Usually this drug is administered as an initial loading dose of 50 mg orally.

Meclizine: This drug is one of the most useful antiemetics to prevent / treat nausea and vomiting associated with vertigo of vestibular origin. It has a slower onset and a longer duration of action (24 hours). For vertigo the usual dose administered in adults is 25 - 100 mg daily in divided doses. Side effects of this drug include: drowsiness, blurred vision, drowsiness.

Promethazine: This drug has pronounced antihistaminic activity in addition to its strong central cholinergic blocking activity. It is effective in the treatment of vertigo and motion sickness. It is administered usually in doses of 25 mg every 4 to 6 hours. One major advantage of this drug is that it can be administered rectally, when severe vomiting prevents its effective oral administration. Most common side effect of this drug is sedation.

Maintenance therapy:

The goal of maintenance therapy is

1. To prevent acute attacks of vertigo

2. To maintain hearing in Meniere's disease

This therapy usually includes dietary modifications combined with pharmacological intervention.

Dietary modifications: The mainstay of diet modifications is to reduce sodium intake. A very low sodium intake or low sodium diet is usually recommended. A strict low sodium diet means a daily allowance of 1500 mg. This is a very stringent diet and patients find it very difficult to comply with this diet. A more practical approach would be to advise the patient to avoid excessively salty food. Restrictions are also imposed on the intake of caffeine, nicotine and alcohol.

Diuretics:

The use of diuretics in the maintenance therapy is based on the supposition that these drugs can alter the fluid balance of inner ear, leading to a depletion of endolymph and a correction of hydrops. In 1934 Furstenburg demonstrated that the symptoms of Meniere's disease were due to retention of sodium. He went on to recommend a low sodium diet / use of diuretics to control Meniere's disease. Boles in 1975 demonstrated that most patients had their vertigo controlled with an 800 - 1000 mg of sodium diet / day.

Hydrochlorothiazide: This diuretic causes natriuresis and kaliuresis by blocking sodium reabsorption in the loop of Henle. Potassium supplementation is required in patients using this drug. Side effects of this drug include: hypokalemia, hyperglycemia, hypotension, and hyperuricemia. It is usually administered as 50 mg tabs orally / day in adults. Potassium supplements are usually required in these patients.

Dyazide: Is potassium sparing diuretic. It can be conveniently administered as a single daily dose.

Frusemide: This is a loop diuretic. It is a very potent diuretic. It can cause electrolyte and volume depletion more rapidly than other diuretics. It usually causes hypokalemia. Usual adult dose is 10 - 80 mg/day. The duration of action lasts for about 4 hours.

Amiloride: This is potassium sparing diuretic acting on the distal tube of Henle. Its diuretic potency is highly limited. It is usually used in combination with other diuretics in order to minimize potassium loss.

Carbonic anhydrase inhibitors:

Acetazolamide: Is a carbonic anhydrase inhibitor. It causes a decrease in the sodium - hydrogen exchange in the renal tubule inducing diuresis.

Methazolamide: Is another carbonic anhydrase inhibitor shown to be effective in controlling symptoms of Meniere's disease. This drug is usually administered in doses of 50 mg / day, 5 days a week for 3 months.

Medical ablative therapy:

Aminoglycosides: Ototoxic effects of aminoglycosides are well documented. Streptomycin and gentamycin are predominantly vestibulotoxic. Intramuscular injections of streptomycin administered twice daily for periods of days to weeks have been used in patients with debilitating bilateral disease / unilateral disease in the only hearing ear. Complete ablation causes disabling oscillopsia. Many authors have suggested lower doses and fewer injections to achieve partial ablation, thereby reducing the incidence of severe ataxia. Currently the recommended daily dose is 1 g of streptomycin intramuscularly 5 days a week until vestibular ablation occurs as manifested by absence of ice water caloric test. Intratympanic injections of these drugs have also been used with success.

Vasodilators:

The use of vasodilators is based on the idea that Meniere's disease results from ischemia of the stria vascularis. Betahistine has been used with varying degrees of success. This drug can be used for short term control of vertigo and for maintenance therapy.

Nicotinic acid is another vasodilator which when administered 30 minutes before meals in doses of 50 - 400 mg helps in resolving the acute crisis associated with Meniere's disease.

Calcium channel blockers:

Nimodipine a highly lipophilic drug is very useful in the medical management of Meniere's disease. It readily crosses the blood brain barrier. This drug is useful in patients who have failed diuretic medical therapy.

ACE inhibitors:

These are very effective vasodilators. These drugs block the renin angiotensin aldosterone system. They produce vasodilatation by blocking angiotensin II induced vasoconstriction.

Lipoflavins and vitamins:

Combination of lipoflavins and vitamins have been tried as a management modality with varying degrees of success.

1.

Ménière's disease - Balasubramanian Thiagarajan - Google Books [Internet]. [cited 2011 Dec 29]; Available from:

http://books.google.co.in/books?id=QjUf0cpT_0MC&printsec=frontcover&dq=Menieres+disease+balasubramanian&hl=en&sa=X&ei=Giv8TuHjHca3rAfkobT-Dw&redir_esc=y#v=onepage&q=Menieres%20disease%20balasubramanian&f=false

2. Medical and non invasive therapy for Meniere's disease Greenberg SL [Otolaryngol Clin North Am.](#) 2010 Oct;43(5):1081-90.

Pendred syndrome ¹

Introduction:

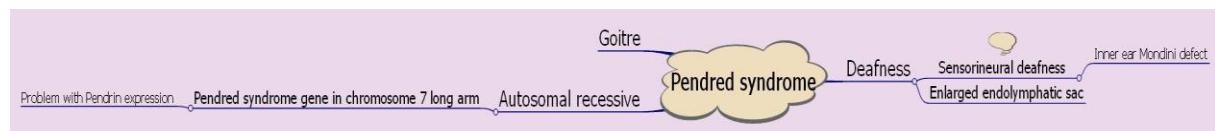
Pendred syndrome was first described in 1896 by Vaughan Pendred.

This syndrome is characterized by:

1. Severe degree of sensorineural hearing loss
2. Goitre.

This condition is characterized by abnormal PS gene in the long arm of chromosome 7. This PS gene is responsible for secretion of a protein called Pendrin.

Pendrin is an anionic exchanger which can cause exchange of chloride, bicarbonate and formate across membranes. Pendrin has been found to be active in thyroid follicular cells. They play a role in transportation of iodide from the cell to the colloid space. This accounts for the presence of goitre in the absence / abnormality of pendrin in these patients.



These patients may manifest with euthyroid state / mild hypothyroid state. Thyroid picture may be highly variable in these patients.

Inner ear findings:

Inner ear shows Mondini type deformity. These patients also show enlarged endolymphatic duct and sac.

In normal conditions, pendrin maintains the ionic exchanges between perilymph and endolymph in the membranous labyrinth which is contained in the bony structure named vestibular aqueduct. If pendrin function is lost, the endolymph volume increases resulting in the enlargement of the membranous labyrinth and of the surrounding bony structures, such as the vestibular aqueduct and the cochlea.

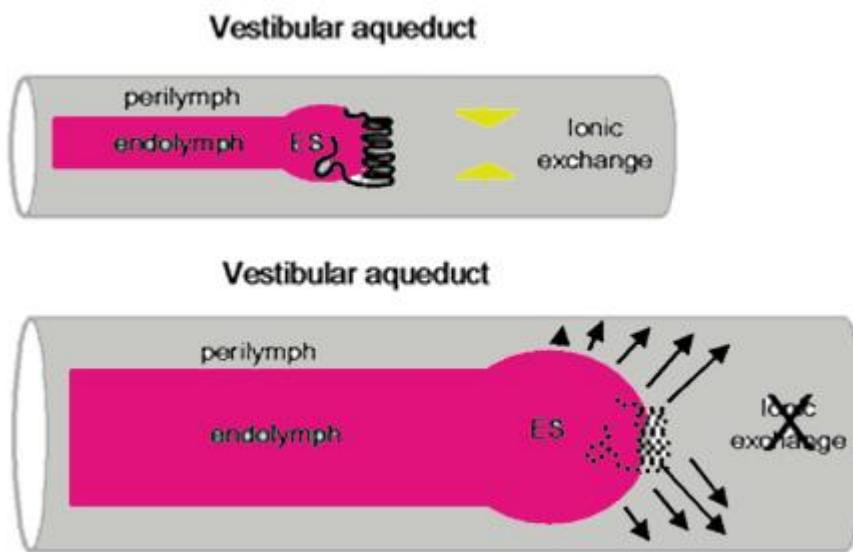
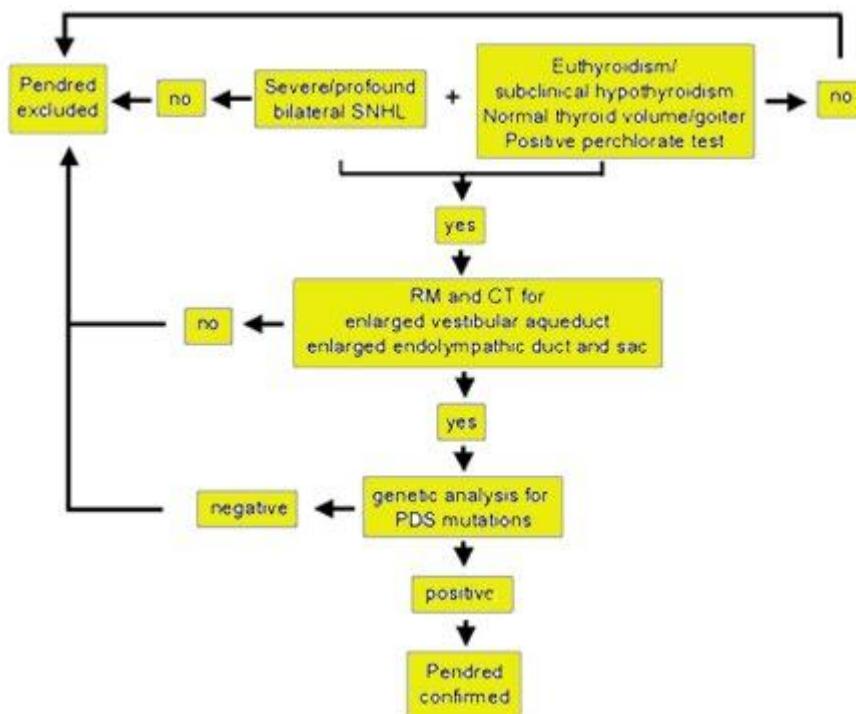


Diagram showing the effects of altered Pendrin in endolymphatic duct / sac.

Vestibular disorder is very rare in this syndrome.

In the absence of inner ear malformations the diagnosis of Pendred syndrome should never be made.

Pendred syndrome should be differentiated from Pendred syndrome like disorders i.e. pseudo pendred syndrome.



Pendrin also play a vital role in the bicarbonate transport in renal tubules. Hence it plays a vital role in the finer electrolyte hemostasis of renal tubules. It also decreases urine chloride excretion. Many of these patients do not manifest with renal problems.

Citation:

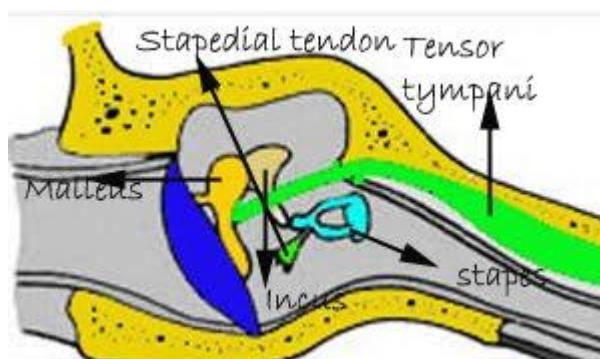
1.
Pendred syndrome - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/pendred-syndrome>

Role of Middle ear muscles ¹

Middle ear houses two important muscles:

1. Tensor tympani
2. Stapedius

Tensor tympani: Originates from the cartilaginous and bony portions of the Eustachian tube. It gets inserted into the handle of malleus after going around the processus cochleariformis. Contraction of this muscle pulls the malleus medially and anteriorly, at right angles to the normal direction of vibration. Contraction of this muscle can be seen as in drawing of the ear drum.



Stapedius: Arises from the pyramid present in the posterior wall of middle ear cavity.

It gets inserted to the neck of stapes.

Contraction of stapedius muscle causes fixation of stapes. It increases the stiffening effects of middle ear conduction mechanism. Contraction of this muscle can reduce transmission by up to 30 dB for frequencies less than 1-2 KHz.

Contraction of these two muscles serves to dampen unwanted resonances in the middle ear system causing spoken words to be heard with clarity.

Stapedius muscle contracts in response to loud sound. The reflex arc of stapedius muscle has 3 - 4

synapses ending in the facial nerve. Since the reflex arc is very small the reaction time is also pretty short i.e. 6-7 milliseconds.

Both these muscles are known to contract in response to non acoustic stimuli also i.e. stimulation of cornea by puff of cold air, vocalisation, touching the skin around the eye etc.

Functions of middle ear muscle reflex:

1. Protection of inner ear from damage due to excess noise. Although the reflex may be pretty slow in protecting the inner ear against sudden noise, it could serve this purpose with longer lasting noise exposures. In fact it has been demonstrated by Zakrisson that patients with Bell's palsy and paralysis of stapedius muscle have a greater incidence of temporary threshold shift when compared with normal controls.

2. Selective attenuation of low frequency sounds emitted from normal body mechanisms like flowing of blood through vessels etc by these muscles have been shown to improve the intelligibility of speech.

Citation:

1.
Role of middle ear muscles - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/role-of-middle-ear-muscles>

Differential diagnosis of cavity in x-ray mastoid

Till recently X ray mastoids was a commonly performed radiological investigation. Here we will be discussing the various causes of cavity seen in x-ray mastoids.

1. Cholesteatomatous cavity: Radiologically this cavity will be surrounded by a rim of sclerosis.
2. Granulation cavity: Cavity will show hair line appearance
3. Operated cavity: Patient will give history of previous mastoid surgery. The margins of the cavity may be irregular and will not show sclerosis
4. Secondaries
5. Multiple myeloma
6. Tuberculosis
7. Eosinophilic granuloma
8. Large antral air cell - usually bilateral

This is a commonly asked question in oral examinations of otolaryngology.

Tests of Eustachian tube function ¹

Functions of Eustachian tube are:

1. Ventilation of middle ear
2. Drainage of middle ear secretions
3. Protection of middle ear from nasopharyngeal commensals and pathogens

For any middle ear surgical procedure to succeed a normal and functioning Eustachian tube is a must.

It is always better to have a clear understanding of the functional status of ET before embarking on tympanoplasty / myringoplasty.

Tests of Eustachian tube function:

Pneumatic otoscopy:

A normal appearing ear drum in otoscopy portends a normal Eustachian tube function. A retracted ear drum indicates a blocked Eustachian tube.

Fluid level seen behind the ear drum indicates secretory otitis media.

A normally moving ear drum on pneumatic otoscopy indicates normal Eustachian tube function.

Valsalva manoeuvre:

This manoeuvre is not popular now. It was first proposed by *Antonio Maria Valsalva* during 17th century. This manoeuvre is performed by exhaling forcibly exhaling against a closed airway. This is a very difficult manoeuvre to perform. To overcome this difficulty a modified Valsalva manoeuvre has been proposed. In this manoeuvre, the patient is made to expire against closed glottis.



Figure showing modified Valsalva manoeuvre being performed.

Toyenbee manoeuvre: This manoeuvre is again used to subjectively / objectively tests Eustachian tube function. The patient is instructed to swallow while pinching both the nostrils. A normally functioning Eustachian tube will cause a popping sound to occur inside the ear. If otoscopic examination is performed simultaneously the ear

drum could be seen moving in and out.

Lowry technique: This is a combination of Valsalva and Toyenbee manoeuvres. This technique involves pinching the nose while attempting to blow and swallow at the same time.

Frenzel Manoeuvre: The patient is asked to pinch the nose closed. The back of the throat is closed (as it happens when one strains to lift a heavy weight). While performing this procedure the patient attempts to vocalize the letter "k". This forces the tongue backwards causing air to be pumped into the Eustachian tube.

Yawning Manoeuvre: Attempting to open the mouth wide as if one is yawning will open the Eustachian tube if it is patent.

Tympanometry:

This test is used to objectively assess Eustachian tube function. Measurement of impedance of middle ear by this procedure will help to assess its function. A normal middle ear pressure is associated with normal Eustachian tube function, while a negative middle ear pressure indicates a blocked Eustachian tube.

Imaging:

CT scan / MRI scan of temporal bone will also help in assessing Eustachian tube function.

Sonotubometry:

Sonotubometry is also known as acoustic tube endoscopy. This method of investigation tests the patency of Eustachian tube

by its ability to conduct sounds from the nasal cavity. A small speaker is placed over the nasal cavity. This speaker is used

to generate sounds at the level of nasal cavity. This sound if the Eustachian tube is patent gets conducted to the middle ear cavity.

A microphone placed at the level of external canal picks up the sound. Eustachian tube function is one of the difficult functions to test clinically, leave alone objectively.

Eustachian tube block is one of the commonest postulated cause for middle ear inflammatory pathologies. Sonotubometry offers a very easy and versatile way of objectively assessing the function of Eustachian tube. This test is also known as acoustic tube endoscopy. Sound usually 8 kHz is generated by a speaker which is placed close to the nasal cavity. This sound will reach the middle ear via the Eustachian tube if it is patent. This sound can easily be recorded by placing a microphone in the external auditory canal.

Four common curves of sonotubometry have been identified by recording sound transmitted

at the level of external auditory canal:

1. Spike type (the most common type 60%)
2. Double peak type (17%)
3. Plateau type (17%)
4. Descendant curve (5%)

In patients with perforated ear drum the function of ET could be assessed by instilling ear drops into the affected ear. If the ET is patent the patient will be able to sense the bitter taste of ear drops in the throat.

Citation:

1.
Tests of eustachean tube function - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/tests-of-eustachean-tube-functin>

Vestibular neuronitis

Definition:

Vestibular neuronitis is characterized by acute onset of severe vertigo with no other signs / symptoms relating to hearing.

Patients with vestibular neuronitis usually have nausea / vomiting without auditory symptoms. Usually these symptoms resolve within a week's time, sometimes the recovery may be more protracted with persistent disequilibrium.

Etiology:

1. Viral infections

2. Labyrinthine ischemia (rare)

Investigations:

Caloric test: has been the traditional gold standard for detecting perpetual vestibular defect.

Recent innovations:

Head impulse test: This test was first developed by Mark Mandala et al in 2005. This test measures the functioning of the lateral canal.

Also known as Head thrust test.

Procedure:

The patient is asked to fix the eyes on their index finger held at an arm's length. The patient's head is moved rapidly to each side (>2000 degrees / sec).

The examiner looks for the presence of nystagmus.

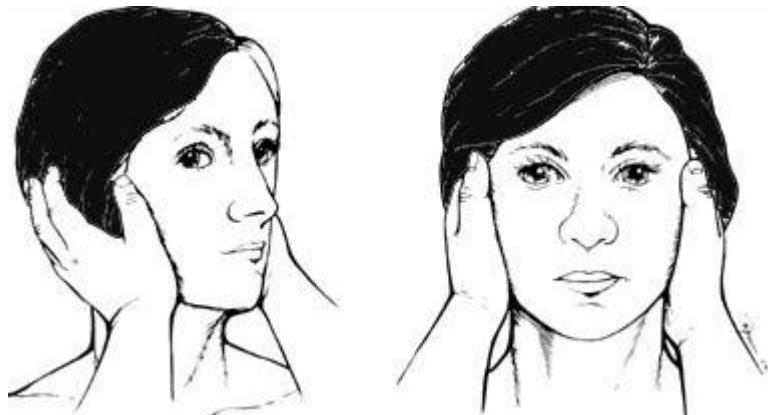


Figure showing Head impulse test being performed

Head heave test:

This test measures the functioning of utricle. This test is performed by heaving the head of the patient rapidly (abrupt, high acceleration, interaural translations (heaves) of about 5 - 10 cms in excursion. The examiner standing in front of the patient should observe for correction saccades.

Vibration induced nystagmus:

This is elicited by using a battery powered vibrator placed over mastoid region for 10 secs, first on one side and then on the other. It is considered to be positive if persistent horizontal torsional nystagmus was evoked for the duration of the stimulus and if the phenomenon is reproducible. The carotid area in the neck is avoided while placing the vibrator.

In case the patient already has spontaneous nystagmus, the test results were considered positive when nystagmus clearly shows an increase in its frequency.



Figure showing the vibrator

Vibrator induced nystagmus depends on direct stimulation of vestibular end organ on both sides because the stimulus is transmitted through the bone to both labyrinth; thus if one side is less responsive the other will predominate. For these tests to be sensitive there must be at least small asymmetry in peripheral vestibular function.

Presence of cerebellar stroke should be ruled out in all these patients as this condition

(pseudovestibular neuronitis) can mimic this condition. MRI should be performed to rule out this condition.

Treatment:

1. Oral methyl prednisolone 60 mg /day tapered during 2 1/2 weeks.
2. Acyclovir 800 mg 4 times a day for 7 days.

Clinical differences between Furuncle / ASOM / Acute mastoiditis

	Furuncle	ASOM	Acute Mastoiditis
H/O cold	No	Yes	Yes
Duration	Short	Short	Long
Tenderness	Over tragus	Nil	Over mastoid
Discharge	Scanty, thick & purulent	Moderate, mucopurulent	Nil / Profuse
Furuncles in other parts	Yes	No	No
Obliteration of retroauricular groove	Yes	No	No
Position of auricle	Pushed forwards	Downwards & outwards	Normal
Otoscopy	Boil in the Ext. canal	Perforated ear drum	Perforated ear drum
X-ray mastoid	Normal	Normal	Cloudy mastoid aircells

Non organic hearing loss ¹

Definition:

Non organic hearing loss can be defined as an apparent hearing loss with no evidence of known disorder or insufficient evidence to explain it.

It is of two types:

1. Psychogenic
2. Malingering

Psychogenic hearing loss:

This includes hearing loss associated with psychological conditions. The patient is not aware that he is simulating deafness.

This is also known as conversion deafness.

Malingering:

Individual is consciously pretending to be deaf to avoid some responsibility / seeking concession even though his hearing may be absolutely normal.

It occurs suddenly and disappears suddenly. It is often associated with mutism when bilateral.

How to identify Malingering?

Quality of voice: Unlike in deaf persons the quality of voice is normal in malingers.

Cochleoauricular / pupillary / palpebral reflexes are normally present in these patients. These reflexes cause twitching of pinna / contraction of palpebral muscles on exposure to loud noise.

Malingering should always be suspected when there is a gross discrepancy between pure tone audiometry and speech audiometry.

Lack of cross over (shadow effect) in pure tone audiometry should cause suspicion.

Tests for malingering:

Chimani Mooss test: This is nothing but a variation of Weber's test. Normally in Weber's test the patient hears the best in the occluded ear. In malingering the patient will not accept to hear better in the occluded ear.

Stenger's test: This test is based on "Stenger's phenomenon". In stenger's phenomenon when a listener is presented with the same type of sound in both ears he /she will hear a single sound, that too only in the ear which it is louder.

Procedure: Two tuning forks with frequency of 512 Hz are kept equidistantly from both ears, one should be able to hear equally well in either side. In malingering say i.e. left ear, even if the tuning fork is moved too close to the left ear, the patient denies that he is hearing in the right side also.

Teal's test: In this test a vibrating tuning fork is applied over the mastoid process of the so called deaf ear, the patient accepts to hear it. Then the patient is blind folded and with a non vibrating fork on the mastoid process, the malingering patient claim's to hear the sound.

Lombard's test: This test is based on "Lombard's principle". This principle says that one raises his / her voice when speaking in noisy environment. While performing this test, the patient is allowed to read a book. Noise is introduced into the ear. The noise is gradually increased till the patient raises his / her voice or stops the process of reading. If there is no change in voice loudness level the patient does not have functional hearing defect.

Erhardt's test: This test is also known as loud voice test. In normal person when the ear is occluded with a finger, it dampens the sound but it can still be heard. Malingerer often denies hearing the sound even when it is loudest.

Stethoscope test: In this test, one ear piece of the stethoscope is closed with wax and used on the side of deafness. The funnel shaped chest piece is used to talk to the patient. The malingerer gets confused and cannot tell whether he is hearing on the right / left side.

Auditory reflex threshold: In normal individuals the stapedial reflex is elicited at 70 - 100 dB. If a malingerer says he is totally deaf and if this reflex is elicited it is suggestive of malingering.

Doerffler Stewart test: This test is based on the fact that persons with normal hearing raise their voice in the presence of background noise. This test can be performed in two ways:

The patient is made to read a passage from a book, while masking noise is fed into the so called deaf ear. In the case of true deafness, the masking noise has no effect on the voice until it reaches the threshold of deafness.

The patient may also be asked to listen to spoken voice instead of reading from a book.

Delayed speech feed back test: In this test the patient is subjected to spoken words whose output is delayed by 200 milliseconds. The level at which it caused difficulty in

speaking is observed. This test is positive in malingers.

Bekesy audiometry: This uses continuous and pulsed tone tracings. The normal graph recorded may be interleaved / continuous tracings below pulsed tone tracings. In patients with non organic hearing loss will have opposite curves - their pulsed tracings are tracked below the continuous tracings. This type of curve is known as Type V Bekesy pattern.

Lengthened off time test: LOT: Conventional Bekesy audiometry uses pulsed tones that are on and off for equal amounts of time (200 milliseconds on and 200 milliseconds off).

The LOT is a test for non organic hearing loss that uses Bekesy audiometry in which the pulsed tones have an off time that is lengthened from 200 ms to 800 ms. In addition the LOT

uses fixed frequency rather than sweep frequency tracings. In this test the continuous tracing is compared to the pulsed tracing that is obtained with a tone that pulses at a rate of 200 ms on and 800 ms off. The LOT increases the degree to which the pulsed tracing falls below the continuous tracing in malingers.

Citation:

1.

Non organic hearing loss by drtbalu - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29]; Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/non-organic-hearing-loss-by-drtbalu>

Visual response audiometry

Synonyms: VRA

Introduction:

This method of hearing assessment is useful in identifying deaf children between 6 months to 2 years of age. This test needs no training of the patient.

The patient is observed for head turn response when sound is emitted by a speaker. When the child turns towards the source of sound, it is rewarded by visual reinforcements like flashing lights or moving toy etc.

Procedure:

As soon as the audiologist gets a reliable head turning response, the intensity of the sound emitted by the speaker is lowered. The lowest sound for which the infant reliably turns its head is noted.

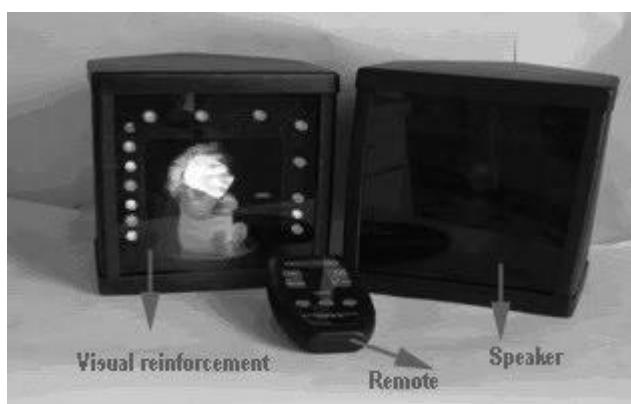
This is infant's VRA threshold. This threshold is not ear specific as sound is emitted via a speaker.

The frequency chosen as stimulus should ideally be within the speech frequency range (500 – 2000 Hz).

Ideally the procedure should not last for more than an hour. The infant should always be provided with breaks in between.

Pitfalls:

1. Fatigability
2. It is not possible to identify unilateral deafness
3. Can only be used as a screening procedure



CROS Hearing aids

Introduction: Cros¹ is an acronym for Contralateral routing of signals. This type of hearing aid is very useful in treating patients with unilateral severe deafness. The principle behind this type of hearing aid is that the good ear is made use of to hear sounds from the bad ear.

In this type of hearing aid the microphone is placed in the bad ear to pick up sounds directed towards the bad ear.

The receiver and amplifier is fixed to the good ear. The sound picked up by the microphone is transmitted to the amplifier fixed to the good ear, it is amplified and projected to the good ear.

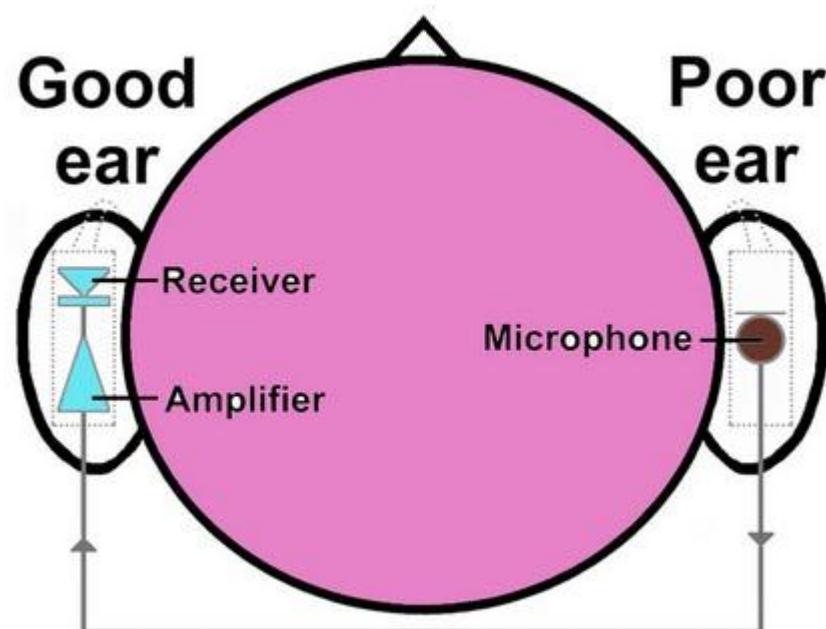
Advantages:

1. Since hearing is in the good ear, amplification need not be maximum
2. There is virtually no feedback
3. Directionality of sound is not lost

Modifications of CROS hearing aids:

BiCROS: This type of aid is used to treat bilateral asymmetrical hearing loss.

SteroCROS: This device is a recent innovation in cros technology. This type of hearing aid restores binaural hearing functionality of ear.



References:

1. Harford, E., Barry, J. (1965). A rehabilitative approach to the problem of unilateral hearing impairment: Contralateral routing of signals (CROS). *J Speech Hear Dis*, 30, 121-138.

1.

- CROS Hearing Aids - drtblau's otolaryngology [Internet]. [cited 2011 Dec 29]; Available from <http://sites.google.com/site/drtbalusotolaryngology/otology/cross-hearing-aids>

Sudden sensorineural hearing loss

Introduction: Sudden sensorineural hearing loss is defined as 20 – 30 dB sensorineural hearing loss in at least 3 contiguous frequencies over a period of less than 3 days. Usually this hearing loss is perceived by the patient suddenly on awakening from overnight sleep. Statistically, this type of deafness constitutes about 1% of all cases of sensorineural hearing loss.

Age incidence: Sudden sensorineural hearing loss may affect persons of any age group. This type of deafness commonly affects patients in their 4th decade.

Associated otological symptoms:

These patients also manifest with associated otological symptoms which include:

1. Tinnitus – 70% of cases
2. Vertigo – 40% of cases

Pure tone audiogram: Shows the following features:

1. Downward sloping – High frequency hearing loss (common)
2. Upward sloping – Low frequency hearing loss
3. U – shaped hearing loss – Mid frequency loss
4. Flat curve – Profound sensorineural hearing loss

Management dilemma:

1. Recovery rates without treatment ranges between 40 – 70%
2. Recurrence rate with / without treatment – 30%
3. Pathophysiology is heterogeneous
4. Incidence is very rare

Causes of sudden sensorineural hearing loss:

1. Idiopathic – 90% cases
2. Non idiopathic – 10% cases

It is very important to rule out acoustic neuroma in all these patients as 4% of these patients may suffer from this disorder. Retrocochlear evaluation is a must in all these patients.

Work up: Should include

1. Pure tone audiometry
2. Impedance audiometry
3. MRI of internal acoustic canal
4. ESR
5. VDRL / FTA – Abs for syphilis
6. Serum tested for autoimmune inner ear disease

Idiopathic sudden sensorineural hearing loss: This condition is probably multifactorial in etiology. This factor goes on to explain the high degree of variability in the therapeutic responses shown by these patients.

Two possible theories have been attributed:

1. Circulatory disturbance ¹
2. Inflammatory reaction

It should also be stressed that these two theories are not mutually exclusive. Viral insults can cause direct neural injury, direct vascular structure injury, direct injury to red cells causing micro vascular compromise. In addition to all these responses, it can also precipitate inflammatory chain reaction.

Advocated treatment for sudden sensorineural hearing loss:

1. Steroids
2. Niacin
3. Histamine
4. Lipoflavinoid vitamins
5. Hyperbaric oxygen therapy ^{3,4}

It has been documented that patients with mid frequency hearing loss had complete recovery even without treatment. It has also been shown that 70% of patients with severe sensorineural hearing loss did not respond to therapy.

Patients with moderately severe sensorineural hearing loss responded to steroid therapy. It has been said that they lie in the “Steroid effective zone”. It is important to identify patients belonging to this group as they could benefit from steroid administration.

Steroid regimen: ²

Currently accepted steroid regimen is oral steroid therapy. Prednisolone is administered in doses of 1mg / kg body weight / day for 7 – 10 days. This is followed by a rapid taper of dose. It has also been said that low dose of steroids are ineffective.

Intratympanic steroid administration:⁵

Advantages include:

1. Drug is directly administered into the site of action
2. It diffuses through round window membrane into the inner ear
3. Since inflammation / autoimmunity have been suspected to the causes, steroids could play a role.
4. There is virtually no systemic side effects

Spinal needle is used for instilling drugs directly into the middle ear cavity. Needle Myringotomy is performed at two different sites in the ear drum. Both these holes should be placed in the antero inferior quadrant. They should not be close enough to avoid the complication of causing a large perforation. Drug is instilled through the second hole and the air from the middle ear cavity could be seen bubbling out through the superior opening on being displaced. Dexamethasone preferably in concentration of 10 mg/cc is used. 1ml of it is mixed with 0.1 ml of 1% xylocaine. About 0.5 ml of this solution is injected into the middle ear cavity. Injections are administered twice a week for a period of 6 weeks. Introduction of Silverstein microwick has made the job little bit simpler.

References:

1. Rudack C, Langer C, Stoll W, Rust S, Walter M. Vascular risk factors in sudden hearing loss. *Thromb Haemost*. Mar 2006;95(3):454-61
2. [Best Evidence] Wei BP, Mubiru S, O'Leary S. Steroids for idiopathic sudden sensorineural hearing loss. *Cochrane Database Syst Rev*. Jan 25 2006;CD003998.
3. Ni Y, Zhao X. [Carbogen combined with drugs in the treatment of sudden deafness]. *Lin Chuang Er Bi Yan Hou Ke Za Zhi*. Jul 2004;18(7):414-5.
4. http://www.drtbalu.co.in/hyper_oxy.html
5. http://www.drtbalu.co.in/tinni_mangmnt.html

Citations:

1. Sudden sensorineural hearing loss by drtbalu - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/sudden-sensorineural-hearing-loss-by-drtbalu>

Shambaugh's criteria to identify patients with sensorineural hearing loss due to otosclerosis

Shambaugh's criteria to identify patients suffering from Sensorineural hearing loss due to otosclerosis:

1. Schwartze sign in either ear
2. Family history of otosclerosis
3. Unilateral conductive hearing loss consistent with otosclerosis and bilateral symmetric snhl
4. Audiogram with a flat / "cookie - bite" curve with excellent speech discrimination
5. Progressive pure cochlear loss beginning at the usual age of onset for otosclerosis
6. CT scan showing demineralization of the cochlea typical for otosclerosis
7. Stapedial reflex demonstrating the biphasic "on-off effect" seen before stapedial fixation

History of Mastoidectomy

Introduction: “One who ignores history would do so at his peril, to be condemned to repeat the same mistakes”. A study of history of mastoid surgery and its instrumentation is important in a sense that they are the tombstones to our success today. Eighteenth century is characterized by advancement in instrument designs and sterilization techniques. Heat resistant metals were used to manufacture surgical instruments as they had to withstand extremely high sterilization temperatures. Our forefathers of 18th century were great innovators and to their credit even now majority of mastoid instruments in use were conceived and designed by them.

Mastoidectomy during different eras:

The art and craft of Mastoidectomy has evolved during the past 200 years. The process of this evolution can be studied under three different eras i.e.:

1. Era of trepan (18th century)
2. Era of chisel & gouge (Early 19th century)
3. Era of electrical drill (20th century)

Era of Trepan:

Trephination was performed to let out pus. This was extensively practiced during the 18th century to let out pus from skull bones. The first successful trephination of mastoid cavity was performed by Ambroise Pare during 16th century. Younger during 17th century devised a hand Trepan which he used extensively to perform this procedure. A hand held trepan was commonly used during this period. The cutting head of trepan used could be circular (to cut a circular piece of bone), exfoliative head (to shed the superficial layer of bone), and perforative head (used to make a hole in the bone). In 1736 Jean Louis Petit performed the first mastoid opening for a patient with mastoid abscess. His main aim was to create a hole through which pus from the mastoid cavity can drain. While using a Trepan it should be dipped in cold water often to reduce heat generated during the procedure.

In 1776 Jasser used a trocar to open up the mastoid cavity. He used the nozzle of a syringe to aspirate the contents from the mastoid cavity. This surgical procedure hence was aptly named as “Jasser procedure”. The term “trocar” has its origin in French language. “Toris – quarts” is a French word to describe an instrument with three cutting sides used to make a hole. American otologist Fredreik White described this era of mastoid surgery as an experimental one. This experimental era proved that the concept of opening up the mastoid cavity and draining the secretions is a possibility. The instrumentation was of

course woefully inadequate. The first catalogue of surgical instruments published in 1860's mentioned the various surgical and dental instruments in use. Mastoid instrumentation of course did not find a place in that catalogue.

Chisel & Gouge period: This period was characterized by the introduction of general anesthesia which facilitated a surgeon to operate leisurely on a patient. It was Amedee Forget a French surgeon who used a mallet and gouge to open the mastoid cavity and drain the accumulated pus. He performed this surgery during 1860.

Modern mastoid surgery was pioneered by the German otologist Schwartze during 1873. He and his assistant Adolf Eysell abandoned the use of Trepan in favour of chisel and gouge. He popularized Chisel and gouge as he was convinced that it was the safest way to open up the mastoid antrum. His assistant had drawn up detailed illustrations of the various types of chisel and gouges used in this procedure. Buck introduced the small curette that could be used to widen the aditus. He also advocated continuous chiseling of the hard mastoid cortex till the soft bone is reached which could be curetted out rather easily using curettes of varying sizes.

Initially Volkmann sharp edged spoons were used as curette. Samuel Kopetzky, American otologist advised that one should become dexterous and elegant with the use of a set of instruments. Newer instruments (design wise) should be introduced only when they have distinct advantages over the tried out older ones. This observation holds good even today.

Electrically driven drill period: "Modern era Mastoidectomy"

Electrically driven drills were used to manage dental caries even way back in 1882. It was William McEwen who drew the attention of the world to this unique device. He believed that the safest instrument that can be used to drill the mastoid antrum is the rotating burr. It had better control and uniform rotator cutting ability. The size of the burr bits can vary according to the area of surgery. It was Julius Lempert in 1922 who really popularized the use of electrically driven drill in ear surgeries. William House introduced the suction irrigation system and retractors in mastoid surgery. He observed that while performing ear surgeries a surgeon needs to keep both hands useful.

Holmgren introduced the operating microscope which really made Mastoidectomy totally a safe procedure.

Middle ear Risk Index MERI

Introduction:

The term Middle ear risk index is used to predict the success rate of middle ear reconstruction procedures. For accurate prediction of the surgical results of middle ear ossiculoplasty the status of middle ear and its ossicles must be ascertained. Austine Kartush classification has been used as a method to define the pre reconstruction ossicular status.

Austine Kartush classification:

This classification uses middle ear ossicular status. Four different groups have been identified:

Group A - Malleus and stapes present (commonly seen status) because of precarious vascularity of incus

Group B - Malleus and foot plate of stapes present

Group C - Malleus absent and stapes present

Group D - Malleus and stapes suprastructure absent

Kartush added three more classes as a modification of this scheme to include ossicular fixity even when all three ossicles are present.

O - Intact ossicular chain

E - Ossicular head fixation

F - Stapes fixation

Middle ear risk index includes:

1. Austin Kartush classification of ossicular defects
2. Ear drum perforation
3. Cholesteatoma
4. Belluci classification

Weightage is given to these 4 parameters to arrive at the middle ear risk index.

Belluci classification uses otorrohea as an index.

Otorrohea	Risk value
Dry ear	0
Occasionally wet	1
Persistently wet	2
Persistently wet with cleft palate	3

Perforation if present adds a value of 1 to the risk index

Cholesteatoma if present adds a value of 2 to the risk index

Austin Kartush classification

1. $M+I+S = 0$

2. $M+S=1$

3. $M-S+ = 3$

4. $M-S- = 4$

5. Ossicular head fixation = 2

6. Stapes fixation = 3

Presence of effusion / granulation in the middle ear adds 2 to the risk index

History of previous surgery adds 2 to the risk factor

History of smoking adds another 2 to the risk factor

Totalling all these factors adds to the middle ear risk index

Citation:

1.
Middle ear risk index (MERI) - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/middle-ear-risk-index-meri>

Dorello's canal

This canal was first described by Gruber in 1859 as an osteo fibrous canal situated at the petrous apex containing the abducent nerve.

This canal is named after the famous Italian anatomist Primo Dorello who proved the existence of this canal after a series of meticulous dissections.

It is found close to the petrous apex. It lies below Gruber's ligament (petrosphenoidal ligament) between the petrous apex and clivus. In majority of cases it is found at the medial most end of petrous ridge at the confluence of inferior petrosal sinus, basal sinus and posterior end of cavernous sinus.

Gruber and Dorello described this canal as the anatomic region between petrous apex and petrosphenoidal ligament. They also included inferior petrosal sinus along with 6th cranial nerve as contents of this canal.

Nathan defined this canal as a little space limited by the apex of the petrous bone, petroclinoid ligament and dorsum sellae. Dolenc believed that this canal extended from the entrance of 6th cranial nerve into the interdural space, up to its entrance into the cavernous sinus.

Involvement of air cells around petrous apex due to mastoid infections can lead to oedema of abducent nerve which lies within this canal.

Facial recess

Introduction:

Is defined as an aerated extension posterior superior portion of the middle ear cavity medial to the tympanic annulus and lateral to the fallopian canal.

Boundaries:

Medial – Facial nerve

Lateral – Tympanic annulus

Superior – Incus buttress (near the short process of incus)

Running through the wall between these two structures with varying degrees of obliquity is the chorda tympani nerve. Chorda tympani nerve always run medial to the tympanic membrane.

Drilling in this area between the facial nerve and annulus in the angle formed by the chorda tympani nerve leads into the middle ear cavity. This surgical approach to the middle ear cavity is known as facial recess approach.

Uses of facial recess approach:

1. Used to reach hypotympanum of middle ear
2. Used to place cochlear implant electrode into the cochlea via the round window.
3. Horizontal portion of facial nerve can be accessed via this approach. Hence this approach can be used to perform decompression of horizontal division of facial nerve.

Occasionally cholesteatoma of middle ear cavity can invade the mastoid antrum without involving the aditus. It has been hypothesized that drilling this area can provide additional avenue for mastoid aeration.

Land marks used to identify this region:

1. External genu of facial nerve medially
2. Fossa incudes superiorly
3. Chorda tympani laterally
4. Tympanic membrane anteriorly and laterally.

Trautman's triangle

This is a triangular space bounded by –

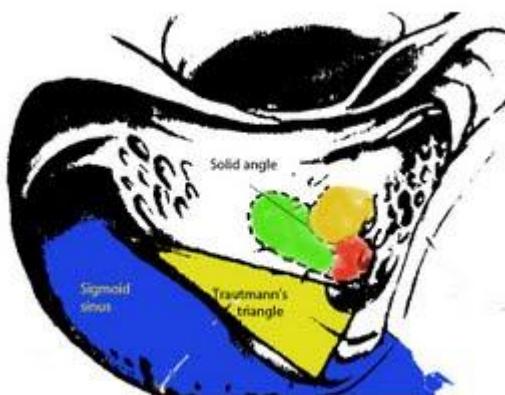
Bony labyrinth anteriorly

Sigmoid sinus posteriorly

Dura containing superior petrosal sinus superiorly.

This triangle is a potential weak spot through which infections of temporal bone may traverse and affect cerebellum. Extra dural abscess involving the posterior cranial fossa is also possible when thin bone in this triangle gets breached in infections / cholesteatoma involving mastoid cavity. Since bone in this area is rather thin it can be drilled out to enter into the posterior cranial fossa. This can be used as an approach to posterior cranial fossa lesions.

The size of this triangle is highly variable depending on the size of the sigmoid sinus. A large sigmoid sinus reduces the size of this triangle and also increases the angulation of the superior petrosal sinus with it. This impedes the venous drainage predisposing to the development of endolymphatic hydrops.



Labyrinthitis

Introduction: This is defined as an inflammatory disorder involving the inner ear / labyrinth. Clinically this condition causes disturbances of balance and hearing of varying degrees in the involved ear.

Causes:

1. Bacterial infections
2. Viral infections
3. Autoimmune causes
4. Vascular ischemic causes

Pathophysiology:

Anatomically labyrinth is composed of an outer osseous framework surrounding the delicate membranous labyrinth which contains the peripheral end organs of hearing and balance.

Membranous labyrinth includes:

1. Utricle
2. Saccule
3. Semicircular canals
4. Cochlea

The labyrinth lies within the petrous portion of temporal bone. It communicates with the middle ear via the oval and round windows.

Infecting organism may find their way into the inner ear via:

1. Pre-existing fractures

2. Oval window
3. Round window
4. Congenital dehiscence involving the bony labyrinth

Viral labyrinthitis: Is characterized by sudden unilateral loss of hearing and equilibrium. Vertigo is usually incapacitating and associated with vomiting. These patients are bed ridden. Vertigo usually subsides within 4-6 weeks. Hearing loss is confined to high frequencies and is sensorineural in nature. An attack of upper respiratory tract infection precedes the development of labyrinthitis. This condition should not be compared with vestibular neuronitis which involves only the vestibular nerves and spares the cochlear component. Varicella Zoster oticus is an unique form of viral labyrinthitis caused by reactivation of dormant varicella zoster virus. This reactivated virus is known to attack spiral ganglion.

Common viral causes of labyrinthitis:

1. Mumps
2. Measles
3. Rubella (congenital labyrinthitis)
4. Cytomegalovirus

Bacterial labyrinthitis: can be potentially caused by meningitis / otitis media. This could be caused by direct invasion of membranous labyrinth by the infecting organism (suppurative labyrinthitis) causing permanent destruction of vestibular and cochlear end organs. In patients with meningitis spread of infections can be bilateral since infections can travel via the CSF and involve the inner ear fluids through the internal acoustic meatus / cochlear aqueduct. Bacterial infections involving the middle ear cavity can enter the labyrinth via erosion of lateral canal which is commonly seen in patients with cholesteatoma. Treatment is usually directed against infecting organism and supportive therapy. Suppurative labyrinthitis is usually followed by labyrinthitis ossificans where the whole of the membranous labyrinth gets ossified. Labyrinthitis ossificans indicates a permanently dead labyrinth.

1. Common bacterial causes of labyrinthitis include:

1. S. pneumoniae

2. *Haemophilus influenza*

3. *Streptococcus*

4. *Staphylococcus*

5. *Neisseria*

6. *Bacteroids*

7. *Proteus*

8. *Moraxella catarrhalis*

Serous labyrinthitis:

This is a potentially reversible disorder caused by diffusion of bacterial toxins into the inner ear via the inflamed round window membrane. Studies have shown that the permeability of the round window membrane is increased when there is inflammation. This may cause diffusion of bacterial toxins and immune mediators into the inner ear causing transient impairment of the inner ear functions.

Autoimmune labyrinthitis:

This is an uncommon cause of sensorineural hearing loss. This may be caused by localized / general autoimmune reactions. Examples of general autoimmune disorders causing labyrinthitis include Wegener's granulomatosis and polyarteritis nodosa.

Citation:

1.

Labyrinthitis - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/labyrinthitis>

Cerumen

This is also known as ear wax. This is a yellowish waxy substance secreted in the external auditory canal of humans. It protects the skin lining of the external auditory canal from excessive moisture. It also protects the external canal from bacteria, fungi and insects.

Humans are known to secrete two types of cerumen:

1. Soft and moist
2. Firm and dry

Persons secreting soft and moist type of ear wax have no problem due to its accumulation. It can easily be extruded by the normal cleansing mechanism of the external auditory canal. This difference in wax secretion has been traced to alterations in C11 gene. Persons secreting firm and dry wax are more prone for impaction of cerumen. Impaction of cerumen causes conductive hearing loss.

Cerumen is usually produced in the outer third of the cartilaginous portion of the external auditory canal. It is composed of:

1. Viscous secretions from sebaceous glands
2. Less viscous secretions from modified apocrine sweat glands
3. Shed layers of skin

Cerumen has been found to have bacterostatic effect. Excessive occlusion of the external canal due to accumulation of cerumen and desquamated epithelial cells associated with migration defect of the lining epithelium can cause keratosis obturans. This is a painful condition which needs to be treated by removing the mass under anesthesia.

Removal of cerumen can be performed using probes / curettes if the consistency is soft. If cerumen is excessively soft then cotton buds can be used for removal.

Firm cerumen should be lubricated by using ceruminolitics / liquid paraffin to soften it up before attempted removal.

Aural syringing is one of the painless ways of removing accumulated cerumen.

Drugs used in otology and their formulations

Introduction: Various drugs and formulations are used in otological practice. Some of them may be of questionable value from the therapeutic stand point, still it is worthwhile knowing about these formulations. Topical otological preparations are so unique they need to be studied in detail.

Advantages of topical drug use in otology:

1. The drug can be administered right where it is needed
2. First pass metabolism doesn't come into play
3. Drug is delivered in a targeted manner, in adequate doses. Toxicity is not common
4. Chronic inflammatory diseases of middle ear cavity cause a certain amount of fibrosis preventing adequate concentrations of systemically administered drug reaching it.

Ear drops are solutions / suspensions of medicines in solvents like water, glycerol, diluted alcohol, or propylene glycol. These solutions can be instilled into the ear. For these ear drops to be effective sufficient contact time should be provided.

Indications for use of topical ear drops:

1. Bacterial / fungal infections of external auditory canal
2. Chronic suppurative otitis media with a large drum perforation
3. To liquefy accumulated wax in the external auditory canal
4. Myringitis granulosa

If drops are to reach the middle ear in adequate concentration the ear drum perforation should be fairly large. The method of administration of ear drops to reach the middle ear cavity is known as the displacement method. In this method the external ear is made dependent by turning the head to the opposite side, with the chin touching the shoulder. The external auditory canal is filled with ear drops. Pressure is applied to the external ear by alternate pressing of the tragus. This maneuver displaces the air from the middle ear cavity which is duly filled up by the ear drops.

Difference between solution and solvent ear drops:

Solution usually consists of a drug which is dissolved in a solvent where as suspension consists of an insoluble drug distributed in a liquid medium. Some of the ear drops can be used as eye drops also. To facilitate such multi usage certain adjuvant drugs are added to the drops in addition to the active drug. Commonly used adjuvants in such drops which can be used as eye and ear drops include:

5. 0.01% Benzalkonium chloride – This acts as an antifungal agent
6. Sodium metabisulphite – This agent acts as a buffer maintaining the pH of the solution. This strict maintenance of pH prevents easy degradation of the active drug molecule present in the drops. It also minimizes the irritation caused due to application of the drops. It also retards the oxidation of the active drug thereby prolonging the effect of the active drug.
7. Disodium edetate – This is another adjuvant commonly used. It also acts as an excellent buffering agent. This adjuvant drug increases the bactericidal and antifungal activity of Benzalkonium chloride.
8. Steroids – Beclamethazone is the commonly used steroid adjuvant drug in the ear drops for its anti-inflammatory effect.

Some of the local drug preparations to be used in the external auditory canal may be in the form of creams / ointments. These ointments usually contain antibiotics and anti-inflammatory agents in a suitable base like liquid paraffin, wool fat, yellow soft paraffin. Ointments usually have paraffin base. Ointments are very useful in managing dry scaly skin conditions of external auditory canal. Ointment preparations with Lanolin as the base (wool fat) should be marked clearly on the tube because some patients may develop hypersensitivity reaction to this component of the medicine.

Acetic acid ear drops – Acetic acid in concentration of 2% is an excellent antibacterial and antifungal agent. Acetic acid ear drops can be used to treat mild otitis externa. This is commonly used in paediatric age group.

Aluminium acetate ear drops: This is an astringent drug which can be administered as ear drops or by dipping a cotton wick in the drug and inserting the same into the external auditory canal. Astringent belongs to a group of medicine that causes shrinkage of tissue on local application. Shrinkage of tissue is caused due to the hydroscopic effect of the drug. Hence it can be used to reduce oedema involving the external auditory canal. If this drug needs to be used for its astringent effect then it should be administered using a cotton wick. This drug is known to cause deposition of aluminium acetate crystals in the external auditory canal. Hence periodic cleansing of the ear is a must when this drug is used. This drug can be safely used even in pregnant mothers. In fact this is safest drug that can be administered during pregnancy.

Boric acid ear drops: Formerly this drug was used for their bacteriostatic and antifungal effects. It can be used in varying concentrations. Maximum concentration that can be safely used is 5%. This drug gets absorbed via the inflamed skin leading on to systemic toxicity due to the drug.

Antibiotic & steroid ear drops:

Betnesol / prednisolone sodium phosphate can be administered along with antibiotics like gentamycin / neomycin / quinolones. When used in combination with these antibiotics they facilitate better effects due to their anti-inflammatory effects.

Clotrimazole ear drops:

Clotrimazole is a broad spectrum antifungal agent. This drug inhibits ergosterol synthesis by the fungal cell wall. This destroys the fungus. Fungal infections involving the external auditory canal can also be caused due to inappropriate use of steroid ear drops. Administration of clotrimazole can cause burning sensation in the external auditory canal. Patient should be advised to tolerate it.

Ceruminolytic ear drops:

These are the most commonly used ear drops. Drugs belonging to this group include oil / aqueous preparations. These drugs are known to soften the wax facilitating its removal by aural syringing. 0.9% sodium chloride solution can be used as Ceruminolytic agent. 5% sodabicarb solution can also be used as Ceruminolytic agent.

Olive oil / coconut oil / liquid paraffin can also be used as Ceruminolytic agents.

Organic solvents like chlorbutanol / paradichlorobenzene can also be used as solvents, but may cause irritation to meatal skin.

Indications for administration for systemic antibiotics:

9. Acute otitis media
10. Furunculosis of external auditory canal
11. Perichondritis of pinna
12. Acute mastoiditis
13. Malignant otitis externa

Drugs administered systemically include:

14. Amoxycillin
15. Flucloxacillin
16. Ciprofloxacin
17. Penicillins

Drugs used in the management of vertigo:

Betahistine can be used in the management vertigo associated with Meniere's disease. This drug reduces the endolymph pressure by improving microvascular circulation in the striavascularis of the cochlea. It also reduces the vertigenous sensation by inhibiting the firing rate of vestibular nuclei. Betahistine is known to reduce vertigo / tinnitus but does little to improve hearing. It is usually prescribed in doses of 16 mg thrice a day.

This drug should be used with caution in patients with bronchial asthma / peptic ulcer.

Dopamine antagonists: Prochlorperazine belongs to this group. Goes by the popular name Stemetil. It is a dopamine antagonist acting by blocking the chemoreceptor trigger zone. It is less sedating with fewer antimuscarinic effects.

Antihistamines: Drugs belonging to this group acts on H1 receptor at the level of chemoreceptor trigger zone thereby blocking the vomiting centre. Examples of drugs belonging to this group are cinnarizine and cyclizine. Cinnarizine can be used as prophylaxis for migraine in doses of 30 mg three times a day. Cyclizine is useful only during acute attacks and is given in doses of 50 mg thrice a day.

Diuretics in the management of Meniere's disease: Thiazides and acetazolamide can be used in the management of acute symptoms of Meniere's disease. They cause decompression of the endolymphatic sac due to their diuretic effects.

Steroids in the treatment of sudden sensorineural hearing loss: Steroids have been used in the management of sudden sensorineural hearing loss with varying degrees of success. Dosage regimen is as follows:

Prednisolone

60 mg on day I

50 mg on day II

40 mg for following three days

30 mg for subsequent three days

Use of antiviral drugs in otology:

Acyclovir is the classic example of drug belonging to this group. It can be administered in patients with Herpes Zoster oticus. It acts by inhibiting nucleic acid synthesis. It is administered orally in doses of 800 mg five times a day for 5 days. If administered within 72 hours after development of rash it reduces post herpetic neuralgia.

Use of sodium fluoroide in otosclerosis:

Sodium fluoride is used to slow down the development of sensorineural hearing loss in a patient with otosclerosis. It acts due to its enzyme inhibiting activity thereby preventing osteoclastic bone resorption. Usually it is administered in doses of 40 mg per day for a period of 3-6 weeks. This drug is really useful in patients with cochlear otosclerosis. It has propensity to cause gastric irritation and renal damage.

Citation:

1.
Drugs used in otology and their formulations - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29];Available from:
<http://sites.google.com/site/drtbalusotolaryngology/otology/drugs-used-in-otology-and-their-formulations>

Golding wood operation

It is also known as Tympanic neurectomy.

This procedure is performed to decrease salivary flow by interrupting parasympathetic fibers of Jacobson's nerve. It was first performed by Golding Wood hence goes by the term "Golding Wood operation".

Indication:

1. In excessive drooling caused by neurological disorders
2. Frey syndrome following parotid surgery
3. Recurrent inflammation of parotid gland if conventional medical management fails

Procedure:

Can be performed both under local / General anesthesia.

Postero superior tympanomeatal flap is created and carried down up to the level of annulus. The incision should be fashioned in such a way that if need arises it can be extended anteriorly along the inferior canal wall to about 5 o clock position. This extension of incision helps in the evaluation of the inferior most portion of the nerve as it enters through the floor of hypotympanum.

The annulus is dissected off its groove and is pushed forwards to 5 o clock position anteriorly and superiorly up to the short process of malleus. After elevation of the tympanic membrane the jacobson's nerve is identified at the promontory as it crosses from inferior to superior direction. This nerve may be completely exposed or partially covered by bone. The hypotympanic portion of this nerve is searched for its antero inferior branch which should be sought and avulsed. This nerve identified at the promontory should be cleared off its bony covering if present and avulsed completely.

Even though majority of parasympathetic nerve supply to the parotid is via the tympanic nerve, it has been demonstrated that chorda tympani nerve also carries a few parasympathetic fibers.

The decision to avulse chorda tympanic nerve along with Jacobson's nerve is highly controversial as it can lead to troublesome xerostomia.

Complications:

1. Residual perforation of ear drum
2. Damage to middle ear structures
3. Rarely troublesome xerostomia



Citation:

1. Golding Wood operation - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 29]; Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/golding-wood-operation>

Susac syndrome

Introduction:

Susac syndrome was first described by Susac et al in 1979. This syndrome is characterised by rapidly progressing encephalopathy, blindness and hearing loss.

Pathophysiology:

This is actually an endotheliopathy affecting precapillary arterioles. This endotheliopathy could probably be immune mediated. This causes rapid tissue infarction which leads to these problems.

Women are commonly affected than men. Typical vulnerable age group is between 20 – 40.

Clinical features:

1. Severe head ache
2. Rapid dementia
3. Micro infarcts seen in corpus callosum demonstrable in MRI scans
4. Photopsia and black spots due to retinal artery occlusion
5. Scintillating scotoma
6. Rapidly progressive sensori neural hearing loss on both sides
7. Vertigo
8. Nystagmus

Management:

High dose steroid therapy is the main treatment modality.

Intravenous administration of immunoglobulin.

Cyclophosphamide administration.

Rituximab is the currently used drug in the management. This is a monoclonal antibody against CD20 protein. This receptor protein is found on the surface of B lymphocytes.

Vestibular Schwannoma current management trends

Introduction:

Management of vestibular schwannomas has undergone lots of changes during the past decade. Review of published literature exemplifies this fact. Various currently available management modalities to treat this condition are:

1. Observation
2. Stereotactic radiosurgery
3. Microsurgery

Among these three modalities stereotactic radiosurgery is evincing keen interest because of the precision of the procedure and lesser incidence of side effects. Advances in imaging technology have enabled early diagnosis of these lesions. About a decade back the sensitivity of imaging techniques used to identify lesions measuring 30 mm. Recent imaging modalities are accurate enough to identify even lesions measuring less than 10 mm. A stage has reached when surgeons are managing more intracanalicular lesions than ever before.

The current management modality of these tumors focusses on:

Preservation of hearing

Preservation of facial nerve functions.

Observation / Watchful waiting: This modality is preferred in managing patients with small asymptomatic / minimally symptomatic intracanalicular tumors. Since tumor doubling time of these lesions is prolonged (1-2 mm / year) this method warrants a trial. Advantages of this method are preservation of hearing and facial nerve function in these patients. Studies have also revealed that growth rates between intracanalicular and extracanalicular tumors are not significantly different. It is ideal to perform imaging at least twice a year within the first year of diagnosis and once a year from there on.

Positive features that could warrant this management modality include:

1. Excellent speech discrimination scores
2. Growth rate of less than 2.5 mm / year

Microscopic surgery:

This is indicated for small intracanalicular lesions with vestibular symptoms. Amount of tumor growth also is one important factor that could force the hands of a surgeon. Growth rate of more than 3mm / year is an indication for surgical intervention. Hearing can be conserved by using retrosigmoid / middle cranial fossa approach.

Stereotactic radiosurgery:

This is indicated in residual lesions after microscopic excision or rapidly enlarging canalicular lesions. Advantages of radiosurgery include:

1. Hearing preservation
2. Conservation of facial nerve function

Grommet insertion current trends

Introduction:

Myringotomy with grommet insertion was introduced by Poltizer of Vienna in 1868. He used this procedure to manage “Otitis media catarrhalis”. Soon it became the common surgical procedure performed in children.

Indications:

Bluestone and Klein (2004) came out with revised indications for grommet insertion which took into consideration the prevailing antibiotic spectrum.

1. chronic otis media with effusion not responding to antibiotic medication and has persisted for more than 3 months when bilateral or 6 months when unilateral.
2. Recurrent acute otitis media especially when antibiotic prophylaxis fails. The minimum episode frequency should be 3/4 during previous 6 months / 4 or more attacks during previous year.
3. Recurrent episodes of otitis media with effusion in which duration of each episode does not meet the criteria given for chronic otitis media but the cumulative duration is considered to be excessive (6 episodes in the previous year)
4. Suppurative complication is present / suspected. It can be identified if myringotomy is performed.
5. Eustachian tube dysfunction even if the patient doesn't have middle ear effusion. Symptoms are usually fluctuating (dysequilibrium, tinnitus, vertigo, autophony and severe retraction pocket).
6. Otitis barotrauma in order to prevent recurrent episodes.

Problems with Grommet insertion:

This procedure is not without its attendant problems. Common problems include:

1. Segmental atrophy of tympanic membrane
2. Tympanosclerosis
3. Persistent perforation syndrome (rare)

Before treating patients with otitis media with effusion the following factors should be borne in mind.

Pneumatic otoscopy should be used to differentiate otitis media with effusion from acute otitis media.

Duration of symptoms should be carefully documented.

Children with risk for learning / speech problems should be carefully identified.

Hearing should be evaluated in all children who have persistent effusion for more than 3 months.

Grommet insertion can be performed under local anesthesia.

Incision is made in the antero inferior quadrant of ear drum. The incision is given along the direction of radial fibers of the middle layer of ear drum. This causes minimal damage to the radial fibers. It also enables these fibers to hug the grommet in position.

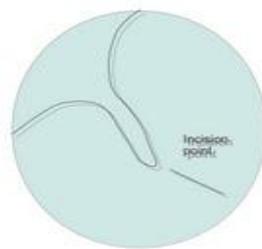


Diagram showing site of insertion of grommet

Retrotympanic recess

Introduction:

The posterior wall of middle ear cavity (Tympanum) is also known as retrotympanum. Important anatomic structures are lodged in this area. This area has assumed significance because of the difficulties encountered in clearing cholesteatoma from this area. This area is so narrow and has lot of crevices; it is very difficult to clear disease from this area.

This area is supposed to contain 4 important recesses. Each of these four recesses could hide cholesteatoma causing the surgeon to leave residual disease which could later recur. Precise knowledge of anatomy of this region is vital for the surgeon who wants to clear disease from this area. The recesses present in the retrotympanic area are:

1. Sinus tympani
2. Lateral tympanic sinus
3. Posterior tympanic sinus
4. Facial recess

Pyramidal eminence is the most prominent anatomical landmark of this area. This eminence holds the pyramidalis muscle. There are other prominences arising from this area projecting in various directions. They include:

1. External: Chordal ridge
2. Inferior: Pyramidal ridge
3. Superior: Suprapyramidal ridge
4. Internal: Ponticulus

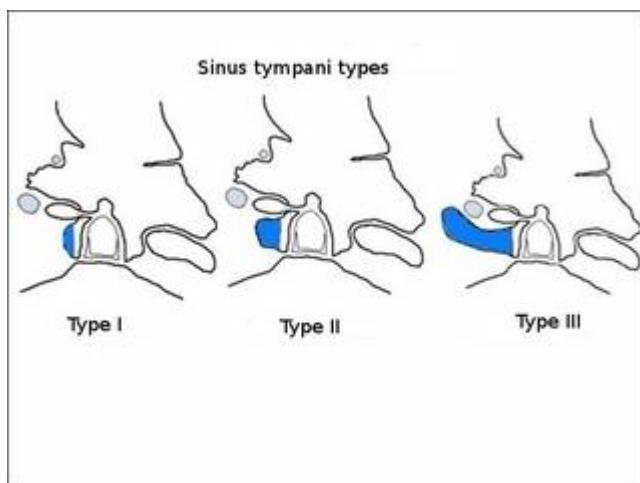
The 4 types of retrotympanic recesses are found under these eminences.

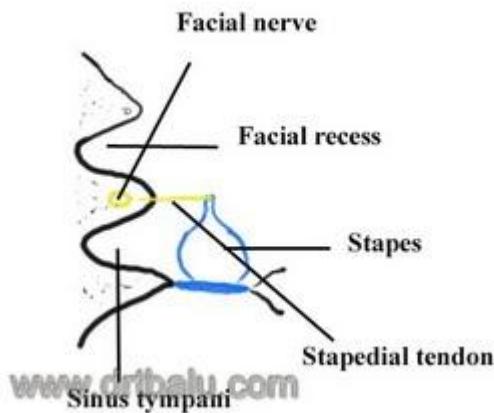
Sinus tympani:

This is the most common and constant depression present in the retro tympanic area. Anatomically this sinus is located at the junction of the lateral and posterior walls of the tympanic cavity. Phylogenetically this recess is considered to be an analogue of bulla tympanica seen in mammals. It lies between ponticulus superiorly and subiculum inferiorly. This recess is bounded by pyramidal ridge externally and promontory internally. Visualization of this area during middle ear surgery proves to be a challenge. During yester years small angled mirrors known as zinne mirrors were used. Now angled telescopes serve this function rather brilliantly. The sinus tympani is known to extend posteriorly up to the round window niche.

Types of sinus tympani:

Sinus tympani has been classified into three types depending on its depth. Note in type III it extends up to the level of lateral semicircular canal.





Lateral tympanic sinus:

Proctor described this sinus in 1969. This sinus lies between three eminences of styloid prominence. These eminences include:

Pyramidal eminence

Styloid eminence

Chordal eminence

Posterior tympanic sinus:

Posterior sinus of middle ear cavity is one of the recently identified anatomical sinus inside the middle ear cavity.

Serial temporal bone dissections have shown that it is present in nearly 90% of dissected bones.

Position: It lies just posterior to the oval window.

Depth: 1mm or less

Width: 1.5 mm or less

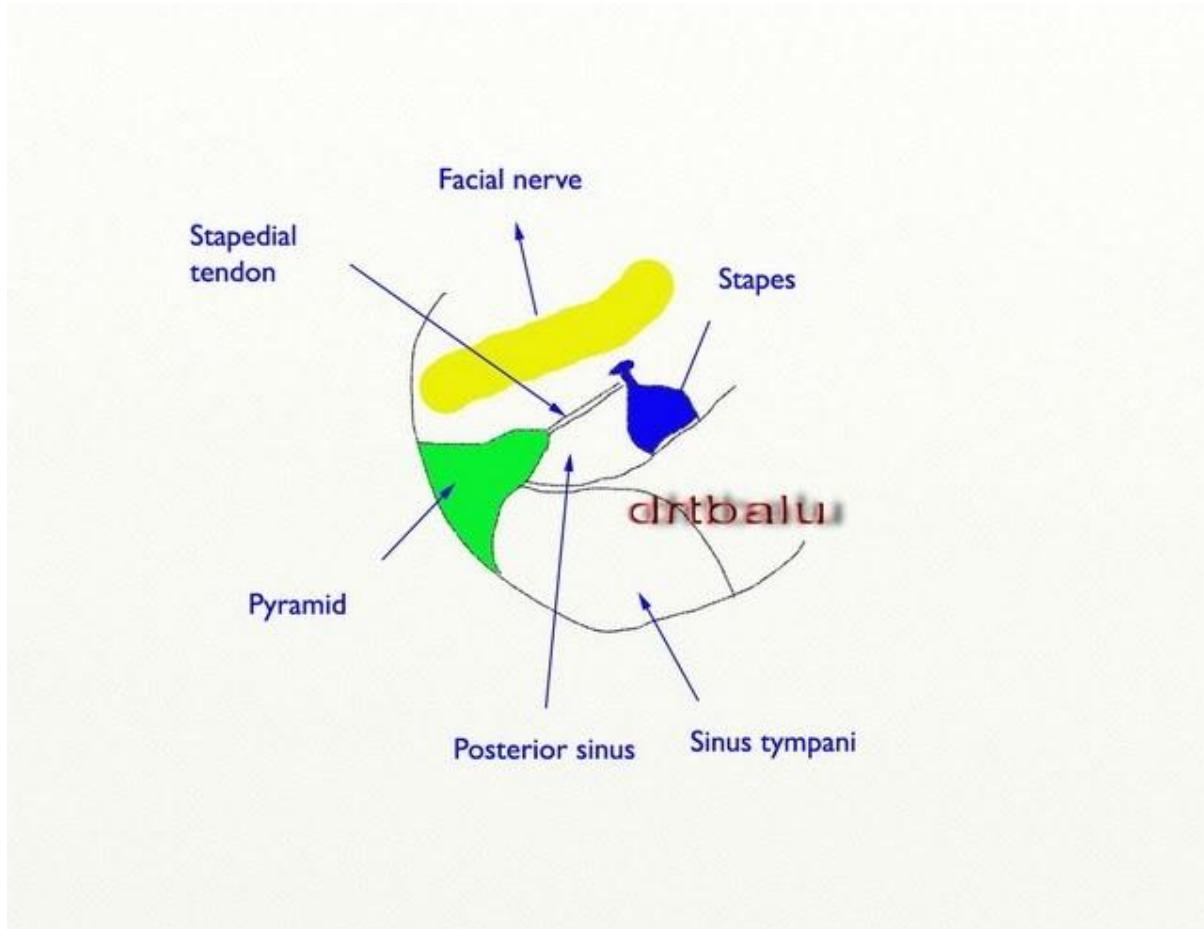
In nearly 60% of dissected specimen a ridge of bone arising from the floor of middle ear

cavity separates it from sinus tympani.

In 8% of dissected specimen, the sinus tympani and posterior sinus merged together to form one confluent sinus.

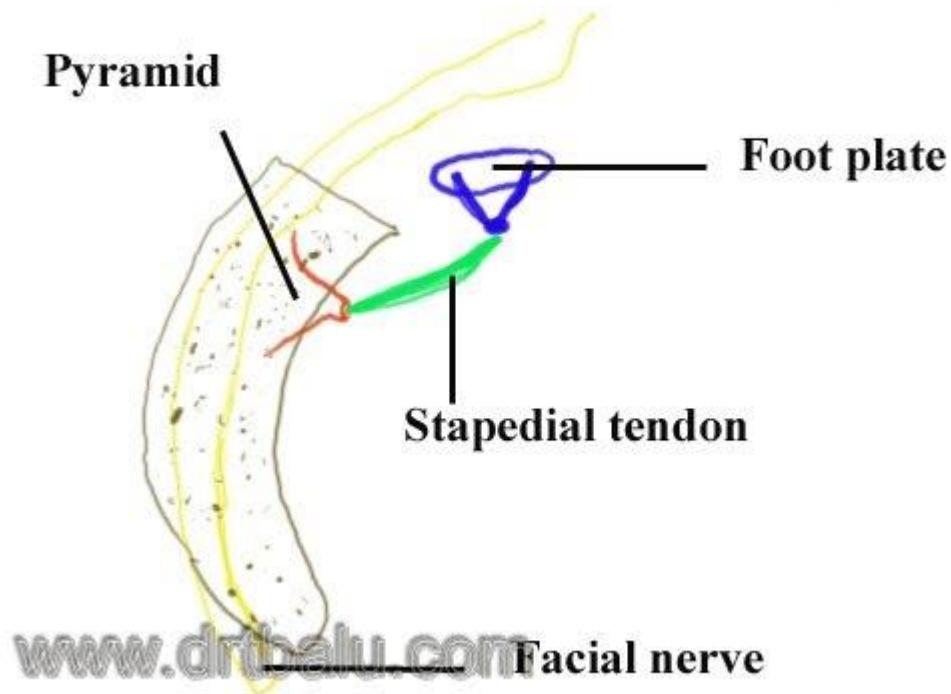
It has been demonstrated that cholesteatoma / granulation tissue may lie within this sinus making removal difficult leading on to residual disease.

Retraction pockets may also occur close to this area.



Facial recess:

This recess lies between the promontory and tympanic annulus. It is bounded medially by the facial nerve and laterally by tympanic annulus. Running between these two structures at varying angulations is the chorda tympani nerve. Chorda tympani nerve always runs medial to the ear drum. Drilling in this area between the facial nerve, annulus and the angle formed by the chorda tympani nerve will lead into the middle ear cavity without causing a breach in the ear drum. This approach is used in cochlear implant surgery to place the electrode in the round window area. Hypotympanum can also be approached through this approach.

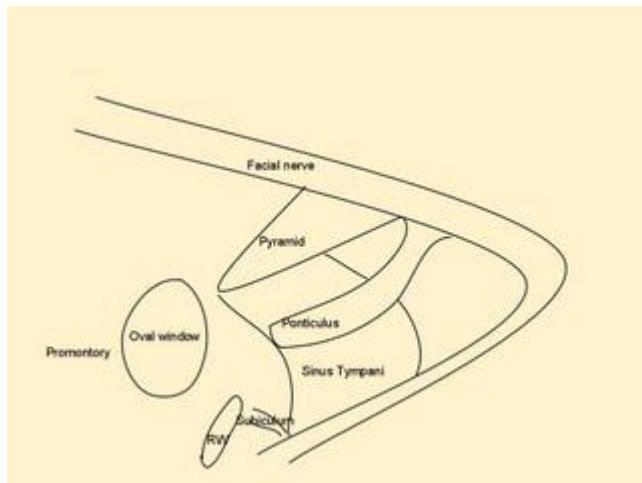


Subiculum:

This is the posterior extension of promontory separating oval and round windows.

Ponticulus:

Rarely a spicule of bone arises from the promontory above the subiculum and runs to the pyramid on the posterior wall of the middle ear cavity. This spicule of bone is known as the ponticulus.



Bibliography:

1.
Retrotympanic Recesses - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 30]; Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/retrotympanic-recesses>

Eosinophilic otitis media

Abstract:

Eosinophilic otitis media is actually a recent introduction. These patients may manifest with sudden hearing loss. There may be associated bronchial asthma and allergic rhinitis. Diagnostic criteria of this condition are rather vague. A review of literature shows that demonstration of eosinophils in the middle ear secretion of these patients could be considered to be pathognomonic of this condition.

Introduction:

Eosinophils are considered to be effectors for allergic reactions. Eosinophilic otitis media¹ is a newly recognised entity causing intractable middle ear pathology. This condition is characterised by excessive accumulation of eosinophils in the middle ear cavity and is associated with persistent middle ear effusion. These patients usually suffer from bronchial asthma. The first description of this condition should be credited to Koch² who first reported some patients with middle ear effusion which contained lots of eosinophils. He also added that these secretions were highly viscous and the middle ear mucosa was pinkish in color. The term eosinophilic otitis media was coined by Tomioka et al³ in 1993.

Pathophysiology:

Pathophysiology of this condition is obviously allergy. These patients commonly had associated allergic rhinitis and bronchial asthma. Eosinophils could have been probably attracted to the middle ear cavity by the presence of IL 5⁴ inside the middle ear cavity.

Features of Eosinophilic otitis media ⁵:

1. Sudden deterioration of hearing
2. Bronchial asthma
3. Allergic rhinitis
4. Intractable otitis media
5. Persistent otorrhoea

Incidence:

Incidence of eosinophilic otitis media is not clearly known. Literature search puts it to be rather common cause of otitis media with effusion.

Management:

Patients diagnosed with this condition should be warned of the possibility of sudden deterioration of hearing.

Administration of systemic / topical steroids ⁶ could be of benefit in these patients.

Antihistamines and leukotriene receptor antagonists can also be used with benefit.

Grommet insertion is indicated in patients with acute sudden hearing loss.

References:

1. Iino Y, Kakizaki K, Katano H, Saigusa H, Kanegasaki S. Eosinophil chemoattractant in middle ear patients with eosinophilic otitis media. *Clin Exp Allergy* 2005;35:1370–6.
2. Koch H. Allergical investigations of chronic otitis. *Acta Otolaryngol* 1947;62(Suppl.):1–201.
3. Tomioka S, Yuasa R, Iino Y. Intractable otitis media in cases with bronchial asthma. Recent advances in otitis media. In: Mogi G, Honjol, Ishii T, Takasaka T, editors. Proceedings of the second extraordinary international symposium on recent advances in otitis media. Amsterdam, New York: Kugler Publications; 1993. p. 183–186.
4. Nonaka M, Fukumoto A, Ozu C, Mokuno E, Baba S, Pawankar R, et al. IL-5 and eotaxin levels in middle ear effusion and blood from asthmatics with otitis media with effusion. *Acta Otolaryngol* 2003;123:383–7.
5. Suzuki H, Matsutani S, Kawase T, Iino Y, Kawauchi H, Gyo K, et al. Epidemiologic surveillance of “eosinophilic otitis media” in Japan. *Otol Jpn* 2004;14:112–7 (In Japanese).
6. Iino Y, Nagamine H, Kakizaki K, Komiya T, Katano H, Saruya S, et al. Effectiveness of instillation of triamcinolone acetonide into middle ear for eosinophilic otitis media associated with bronchial asthma. *Ann Allergy Asthma Immunol* 2006;97:761–6.

Citations:

1. Eosinophilic otitis media a literature review - drtbalu's otolaryngology [Internet]. [cited 2011 Dec 30]; Available from: <http://sites.google.com/site/drtbalusotolaryngology/otology/eosinophilic-otitis-media-a-literature-review>

Ramsay Hunt syndrome

Definition:

Ramsay Hunt syndrome is a disease affecting the external auditory canal associated with the following symptom complexes:

1. Lower motor neuron type of facial nerve palsy
2. Herpetic blisters of the skin of the external auditory canal
3. Otalgia

This syndrome was first described by J. Ramsay Hunt in 1907. He described patients with Otalgia associated with cutaneous and mucosal rashes. He attributed it to the infection of geniculate ganglion by Herpes virus type 3.

Pathophysiology:

The primary pathophysiology is located in the geniculate ganglion of the facial nerve. Geniculate ganglion is found to be affected by Human Herpes virus type 3 i.e. (Varicella zoster virus). Varicella zoster virus has been identified from tears of these patients by polymerase chain reaction. In fact Varicella zoster virus has also been identified from tears of patients with Bell's palsy.

These patients have deep seated pain in the affected ear associated with vertigo, tinnitus, ipsilateral transient hearing loss and lower motor neuron type of facial palsy. These symptoms develop due to involvement of the geniculate ganglion of the facial nerve located near the petrous pyramid portion of the temporal bone. The site of rash varies from patient to patient due to individual variations in the areas supplied by the nervus intermedius of Wrisburg (sensory branch of facial nerve). Rashes may be present in the anterior 2/3 of the tongue, soft palate, external auditory canal and the pinna.

Morbidity / Mortality:

This disease is usually not associated with mortality. It is a self limiting disease, with morbidity due to facial nerve palsy. Complete recovery of the nerve is seen only in 50% of patients as compared to more than 90% in Bell's palsy.

Clinical features:

Patient has deep seated pain in the affected ear. The pain is intermittent in nature, radiating towards the pinna of the ear. There is associated diffuse dull aching background pain. Patients also give history of exposure to Varicella virus infections (chicken pox). The classic Ramsay Hunt syndrome is associated with:

1. Pain in the ear
2. Vertigo and ipsilateral hearing loss
3. Tinnitus
4. Facial palsy (LMN type). Rash or blisters can also be seen along the distribution of nervus intermedius. These herpetic blisters in the external auditory canal may become secondarily infected causing cellulitis.



Picture showing blister in the external auditory canal



Picture of a patient with left sided LMN facial palsy

Investigations:

Basic investigations like blood count, ESR and electrolytes estimation must always be done in these patients.

Virology:

1. Varicella virus the causative agent responsible for this syndrome also causes chicken pox in children

2. Serologic tests for Varicella virus is positive

3. Varicella virus can be isolated and cultured from the fluid extruding from the blisters

4. It can also be detected by PCR on samples of tear fluid from these patients.

5. Audiometry demonstrates sensorineural hearing loss

6. Unilateral caloric weakness may be present on electronystagmography (ENG).

Histology:

The affected ganglia are found to be swollen and inflamed. The inflammatory reaction is lymphocytic in nature. Some of the cells in the ganglia may show evidence of degeneration.

CSF analysis is not indicated in these patients.

Management:

1. Steps towards alleviating pain: Carbamazepine can be prescribed in doses of 400 mg / day in divided doses. Temporary relief of Otic pain in geniculate neuralgia may be achieved by applying a local anesthetic or cocaine to the trigger point, if in the external auditory canal.
2. Corticosteroids and oral acyclovir can be administered. Steroids in the form of prednisolone can be administered orally in doses of 10mg twice a day. Steroids should not be stopped abruptly. The dosage needs to be tapered. Acyclovir can be administered in doses of 800 mg orally 5 times a day.
3. Management of vertigo: can be managed using meclizine in doses of 25 mg orally 4 times a day.
4. Care must be taken to prevent exposure keratitis because of the inability to close the eye lids. The patients must wear protective goggles.

Citation:

1.
Ramsay Hunt Syndrome by drtbalu [Internet]. [cited 2011 Dec 30];Available from:
<http://www.drtbalu.co.in/ramsay.html>

BPPV (Benign Paroxysmal Positional Vertigo)

Benign paroxysmal positional vertigo is the most commonly diagnosed vestibular disorder. This is commonly caused by dysfunction of the posterior semicircular canal. Lateral and superior semicircular canals can also be involved on rare occasions. It is characterized by brief spells of severe vertigo (often lasting for just a few seconds) that are experienced only with specific movements of the head.

History:

This disorder was first described by Barany in 1921. He documented the various components of this disorder as:

1. Nystagmus
2. Fatigability of the nystagmus
3. Vertigo.

He failed to correlate the onset of nystagmus with specific positions of the head.

Dix & Hallpike 1952 described the Dix Hallpike maneuver for eliciting the nystagmus. They also described the unique features of nystagmus accompanying this disorder. These features were 1. Very short latency, 2. Directional features, 3. Brief duration, and 4. Reversibility on returning the patient to a seated position.

Schuknecht postulated that BPPV was caused by loose otoconia from the utricle which in certain positions, displaced the cupula of the posterior canal. (Schuknecht theory). He later modified his theory and proposed that it was due to the deposition of otoconia on the cupula of the posterior semicircular canal. He termed this theory as cupulolithiasis. The cupulolithiasis theory proposes that calcium deposits become embedded on the cupula making the posterior semicircular canal sensitive to gravity.

Hall & Ruby suggested that BPPV could result from deflection of the posterior canal cupula caused by debris within the posterior canal. This theory became known as the canalithiasis theory. In this theory the calcium debris does not become adherent to the cupula but float

freely within the canal. Head movements like looking up, down, or rolling over to the affected ear may result in the displacement of the sludge causing the classic symptoms.

Hall & Ruby described 2 types of BPPV: 1. BPPV with a fatigable nystagmus, where the deposits are freely mobile within the cupula of the posterior canal,

2. BPPV with a non-fatiguing nystagmus where the calcium deposits are fixed on the cupula of the posterior canal.

Typical features of BPPV as described by Hall & Ruby:

1. Canalithiasis mechanism - This explains the latency of the nystagmus as a result of the time needed for motion of the material within the posterior canal to be initiated by the gravity.

2. Duration of the nystagmus - is correlated with the length of time required for the dense material to reach the lowest part of the posterior canal.

3. The vertical (upbeating) and torsional (superior poles of the eye beating towards the lowermost ear). The nystagmus is more vertical when the patient looks away from the lowermost ear, and more torsional when looking towards the lowermost ear.

4. The reversal of nystagmus when the patient returns to the sitting position is due to retrograde movement of material in the lumen of the posterior canal back towards the ampula, resulting in ampulo petal deflection of the cupula.

5. The fatigability of the nystagmus evoked by repeated Dix Hallpike positional testing is explained by dispersion of material within the canal.

Incidence:

BPPV is the most common cause of vertigo constituting 20 - 40% of all patients with peripheral vestibular disease. Mean age of onset ranging between 4th and 5th decades. Women outnumbering men by 2:1.

History: Patient c/o severe vertigo associated with change in head position. Symptoms are always sudden in nature, never lasting more than a minute. The patient may even volunteer provoking postures.

On examination: the classic eye movements associated with Dix Hallpike maneuver is seen.

Dix-Hallpike maneuver: The patient is positioned on the examination table in such a way that when he/she is placed supine, the head extends over the edge. The patient is lowered with the head supported and turned 45 degrees to one or the other side. The eyes are carefully observed; if no abnormal eye movements are seen, the patient is returned to the upright position.

This same maneuver is repeated with the head in the opposite direction and the patient's symptoms are noted.

The pattern of response consists of the following:

1. Nystagmus is a combination of vertical upbeating & rotatory (torsional) beating towards the downward eye. Pure vertical nystagmus is not seen in BPPV.
2. There is often a latency of onset of nystagmus

3. Duration is less than a minute
4. Vertiginous symptoms are invariably seen
5. Nystagmus disappears with repeated testing (fatigability)
6. Symptoms often recur with the nystagmus in opposite direction on return of the head to upright position.

Canalithiasis involving the posterior canal is the commonest cause of BPPV. Posterior canal BPPV may rarely be bilateral, but while testing the head must be positioned in the plane of the posterior canal during testing of unaffected ear otherwise the debris in the affected side can rest against the cupula and stimulate an excitatory nystagmus from the unaffected ear.

Lateral canal BPPV:

Lateral canal has also been identified as the offender in 17 % of cases with BPPV. Lateral canal BPPV can be detected by a variation of Dix Hallpike maneuver. The patient's head is first brought to the supine position resting on the examination table (not hyperextended). The head is then turned rapidly to the right so that the patient's right ear rests on the table. The eye movements of the patient are monitored with Frenzel's glasses for 30 seconds. The patient's head is then turned to the supine position (eyes looking upward) and is then rapidly turned to the left so that the left ear rests on the table. Eye movements are monitored. The nystagmus with lateral canal BPPV is horizontal and may beat toward (geotropic) or away (ageotropic) from the downward ear. It begins with a short latency, increases in magnitude progressively, and is less susceptible to fatigue with repetitive testing than the vertical torsional nystagmus of posterior canal BPPV.

Cupulolithiasis, either alone or in combination with canalithiasis is more likely to be involved in the etiology of lateral canal BPPV than in the case of posterior canal BPPV. If the nystagmus is geotropic, the particles are likely to be in the long arm of the lateral canal

relatively far from the ampulla, if the nystagmus is ageotropic, the particles could be in the long arm relatively close to the ampulla or on the opposite side of the cupula either floating within the endolymph or embedded in the cupula.

Superior canal BPPV: Incidence of superior canal BPPV is very rare.

Standard electrooculography or 2 dimensional video nystagmography devices do not record the typical eye movements associated with BPPV. Thus clinical examination of the patient is of paramount importance.

Management:

Medical:

Repositioning maneuver: Currently BPPV is managed by repositioning maneuvers that, in cases of canalithiasis use gravity to move canalith debris out of the affected semicircular canal and into the vestibule. For posterior canal BPPV the maneuver developed by Epley is effective.

Epley maneuvers - This is performed by placing the head of the patient in the Dix Hallpike position that evokes the vertigo. The posterior canal on the affected side is in the earth vertical plane when the head is in this position. After the cessation of initial nystagmus, the head is rolled through 180 degrees, (this is done in two 90 degree increments, stopping in each position until the nystagmus resolves) to the position in which the offending ear is up. The patient is then brought to the upright sitting position. This procedure is likely to be successful when nystagmus of the same direction continues to be elicited in each of the new position (as the debris continues to move away from the cupula). This maneuver is repeated until no nystagmus is elicited. This is successful in 90 % of cases. Posterior canal BPPV can be converted to lateral canal BPPV during Epley maneuver. The lateral canal BPPV resolves in several days. Drugs are usually not prescribed, but low dose meclizine or calmipose can be given 1 hour before the procedure if the patient is anxious or prone to vomiting.

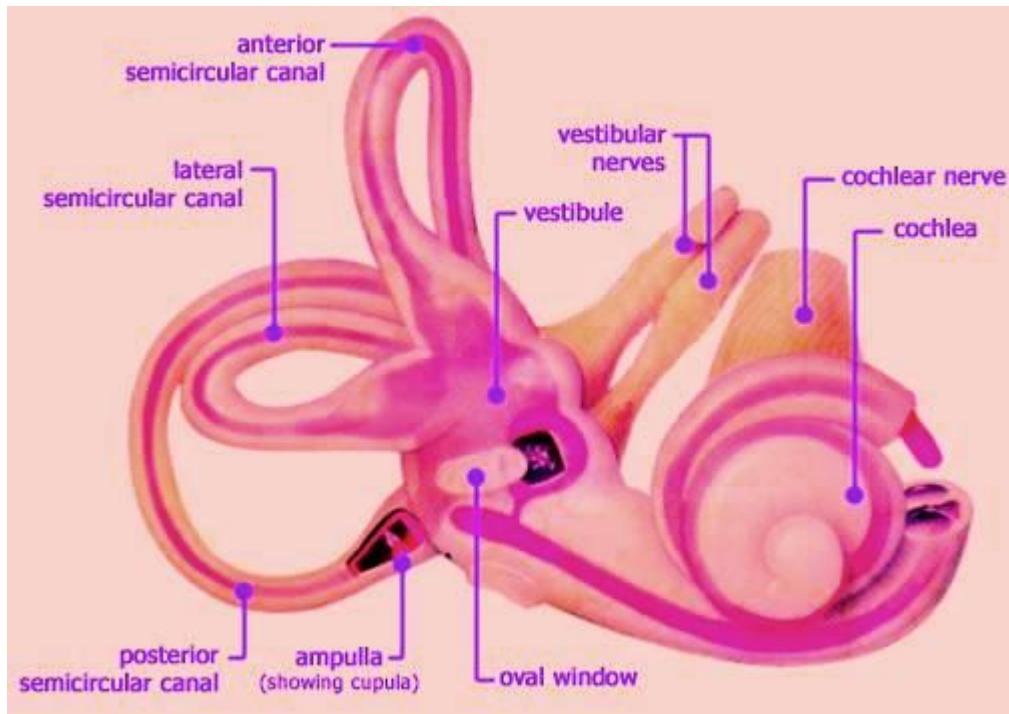


Diagram showing anatomy of semicircular canals

Sermont manoeuvres - is also effective in posterior canal BPPV, but is most difficult to perform and it has no significant advantages over the Epley manuver. This is being described here for the sake of completion. In this manuver the patient is moved quickly in to the position that provokes the vertigo and remains in that position for 4 minutes. The patient is then turned rapidly to the opposite side ear down, and remain in the second position for 4 minutes before slowly getting up.

In both these manoeuvres gravity is the stimulus that move the particles within the canal, so there is no need to turn the head on the body, en bloc movement of the head and body as much as possible is the plan.

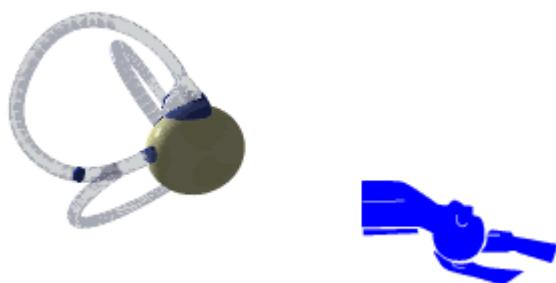


Figure showing repositioning manuver being performed

Vibrator therapy:

Some physicians use a small hand held vibrator over the mastoid to agitate the particles and make it move. This mastoid vibrator is to be avoided in patients with retinal detachment or in patients who may be susceptible to retinal detachment due to high myopia.

After these repositioning maneuvers patients are instructed to avoid bending over and are told to sleep with the head elevated at least 45 degrees for the next couple of days.

Brandt Doroff exercises - can be performed by the patient in the home environment. These exercises are performed in 3 sets / day for 2 weeks.

It is started like this:

Position 1 - The patient must be seated upright on the bed. Then he moves to side lying position (position 2) the head is kept angled upwards about half way. The patient should stay in this position at least for 30 seconds or till the giddiness subsides. If the giddiness does not subside the patient must revert back to position 1. After 30 seconds the procedure is repeated on the opposite side. Most of the patients get relief within a period of 10 seconds.

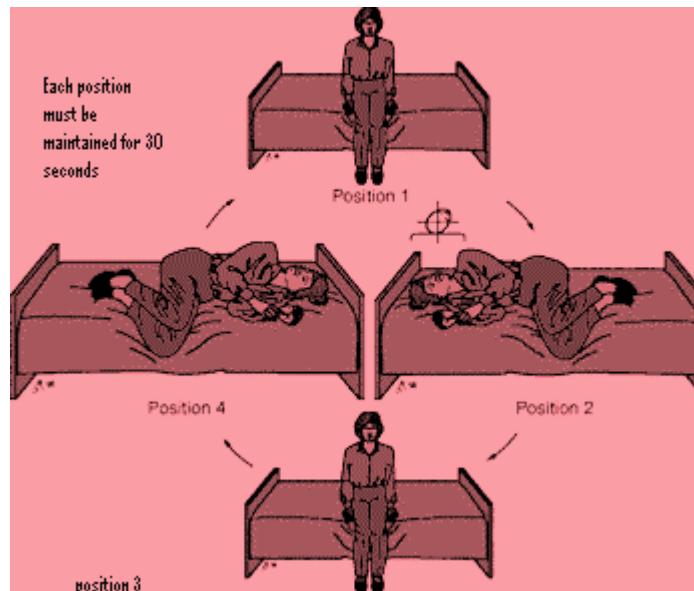


Figure showing Brandt Doroff exercises being performed

Treatment maneuvers for lateral canal BPPV:

In these patients with geotropic nystagmus lying on one side with the affected ear up for 12 hours has been found to be effective.

Surgical management:

Singular neurectomy - is a very demanding procedure. The posterior canal is supplied by singular branch of vestibular nerve. This nerve when preferentially sectioned alleviates the patient's symptom due to posterior canal BPPV.

Posterior canal plugging procedure - is a easier procedure. Through a mastoidectomy incision the labyrinth is exposed. The posterior canal is drilled exposing the membranous portion of the canal. The canal is sealed and packed off thereby preventing the debris from floating. After the procedure the patient may feel slightly giddy. The patient needs to be kept in the hospital till giddiness subsides.

Citation:

1.
BPPV (Benign paroxysmal positional vertigo) by drtbalu [Internet]. [cited 2011 Dec 30]; Available from: <http://www.drtbalu.co.in/bppv.html>

Malignant Otitis externa

Definition: Malignant otitis externa is a inflammatory disorder involving the external auditory canal caused by pseudomonas organism. Majority of these patients are elderly diabetics. This condition is termed as malignant otitis externa because of its propensity to cause complications. Hence the term malignant must not be construed in a histological sense.

History:

1838 - Toulmousch reported the first case of otitis externa

1959 - Meltzer reported a case of pseudomonas osteomyelitis of temporal bone

1968 - Chandler discussed the various clinical features and described it as a distinct clinical entity

The effectiveness of present day antibiotics in the management of this condition should provoke the physicians to abandon the term malignant while describing this condition.

Epidemiology:

The typical patient with malignant otitis externa is an elderly diabetic, with males outnumbering females by twice the number. This could be due to the possibility of males being more prone to secrete wax which are more acidic in nature. Malignant otitis externa is very rare in children; if present it will be associated with malnutrition or HIV infection.

Pathophysiology:

Malignant otitis externa is known to affect the external auditory canal and temporal bone. The causative organism being pseudomonas aeruginosa. These patients are invariably elderly diabetics. This disorder usually begins as otitis externa and progresses to involve the temporal bone. Spread of this disease occurs through the fissures of Santorini and osteo

cartilaginous junction. This disorder could be caused by a combination of poor immune response and peculiar characteristics of the offending microbe.

Immunity is reduced in patients with:

1. Diabetes mellitus
2. Blood cancer
3. HIV infections
4. Patients on anticancer drugs

It should also be remembered that diabetic patients have impaired phagocytosis, poor leukocytic response, and impaired intracellular digestion of bacteria. Diabetic patients secrete wax which has less lysozyme content than normal thereby reducing the effectiveness of wax as an antimicrobial agent.

Pseudomonas aeruginosa is a gram negative aerobe with polar flagella. It is found on the skin. It invariably behaves like an opportunistic pathogen. The pathogenicity of this organism is due to ability to secrete exotoxin and various enzymes like lecithinase, lipase, esterase, protease etc. Since this organism is clothed by a mucoid layer it is resistant to digestion by macrophages.

Clinical features:

The patient gives history of trivial trauma to the ear often by ear buds, followed by pain and swelling involving the external auditory canal. Pain is often the common initial presentation. It is often severe, throbbing and worse during nights. It needs increasing doses of analgesics. On examination granulation tissue may be seen occupying the external canal. It often begins at the bony cartilaginous junction of the external canal. Discharge emanating from the external canal is scanty and foul smelling in nature. When the discharge is foul smelling it indicates the onset of osteomyelitis. Ironically the patient does not have fever or other constitutional symptoms.

Otoscopy: Reveals granulation tissue at the bony cartilaginous junction. The ear drum is usually normal. The external auditory canal skin is soggy and edematous.

Cranial nerve palsies are common when the disease affects the skull base. The facial nerve is the most common nerve affected. As the disease progresses the lower three cranial nerves are affected close to the jugular foramen.

Intracranial complications like meningitis and brain abscess are also known to occur.



Figure showing granulation tissue in the external auditory canal



Picture showing LMN type facial palsy on the left side

Spread of infection:

1. Inferiorly through the stylomastoid foramen to involve the facial nerve.
2. Anteriorly to the parotid
3. Posteriorly to the mastoid and sigmoid sinus
4. Superiorly to the meninges and brain
5. Medially to the sphenoid
6. Spread through vascular channels are also common

Role of imaging:

- * Conventional radiology is of no use.
- * CT scan is useful in assessing bone destruction.
- * MRI is useful in assessing soft tissue involvement.
- * Radionuclide scans with Technetium 99 helps in assessing bone involvement

Imaging algorithm in these patients are:

1. TC99 scan to seek evidence of bone involvement
2. If this is positive CT scan and MRI scan is a must to rule out bone and soft tissue involvement
3. Serial Ga 67 scans to assess the efficacy of treatment modality.

Levenson's criteria for diagnosis of malignant otitis externa:

- * Refractory otitis externa
- * Severe nocturnal otalgia
- * Purulent otorrhoea
- * Granulation tissue in the external canal
- * Growth of *Pseudomonas aeruginosa* from external canal
- * Presence of diabetes and and other immunocompromised state

Staging & classification:

Stage	Ga67	TC99	Extent of Disease
I	+	-	Soft tissue (Necrotising Otitis)
II	+	+	Ear & Mastoid (Skull base osteomyelitis)
III	+	+	Extensive skull base osteomyelitis

Treatment:

Extensive surgical procedures have failed miserably to cure this condition. The role of surgery is confined to only exclusion of malignancy by biopsy. Wound debridement is a possibility in advanced cases.

Medical management:

Carbenicillin, Pipercillin, Ticarcillin can be used. Third and forth generation cephalosporins can be used.

Ciprofloxacin in doses of 1.5 g - 2.5 g /day in divided doses can be administered for a period of 2 weeks.

Gentamycin can also be administered parenterally in doses of 80 mg iv two times a day in adults.

Citation:

- 1.

Malignant Otitis externa by drtbalu [Internet]. [cited 2011 Dec 30];Available from:
http://www.drtbalu.co.in/malig_otitis.html

Acute otitis media¹

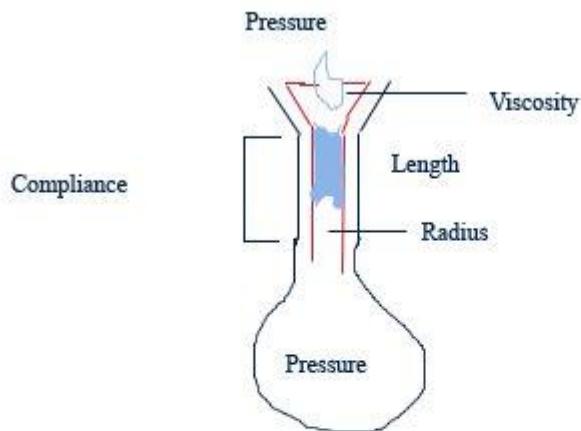
Definition: Acute suppurative otitis media is defined as suppurative infection involving the mucosa of the middle ear cleft. By convention it is termed acute if the infection is less than 3 weeks in duration.

Pathophysiology: Obstruction to the Eustachian tube seems to be the most important antecedent event in the pathophysiology of acute suppurative otitis media. Majority of acute suppurative otitis media is triggered by upper respiratory infections which might find its way into the middle ear cavity through the Eustachian tube orifice. Infections involving the nasopharynx may find its way into the middle ear through the pharyngeal end of Eustachian tube. The infection is initially commonly viral in origin; allergy could also play an important role in the pathogenesis. Later the middle ear mucosa becomes secondarily infected by pathogenic bacteria. The bacteria commonly implicated in this disorder are *S Pneumoniae*, *H. Influenza*, and *M Catarrhalis*.

The majority of otitis media prone children have a patulous Eustachian tube or an hypotonic Eustachian tube. Children with neuromuscular disorders or with abnormalities of the first or second arch have a patulous Eustachian tube leading on to this problem. To become pathogenic the bacteria must become adherent to the mucosa lining the middle ear cavity, this is made possible by prior infection of the middle ear mucosa by viruses.

Flask model explaining the role of Eustachian tube in middle ear infections:

The Eustachian tube, middle ear, and mastoid air cell system can be likened to a flask with a long narrow neck. The mouth of the flask represents the nasopharyngeal end, the narrow neck, the isthmus of the Eustachian tube, and the bulbous portion, the middle ear and mastoid air chamber. The fluid flow through the neck of the flask would be dependent on the pressure at end, the radius and length of the neck, and the viscosity of the liquid. When a small amount of liquid is instilled into the mouth of the flask, liquid flow stops somewhere in the narrow neck owing to capillarity within the neck and the relative positive air pressure that develops in the chamber of the flask.



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Figure explaining the normal Eustachian tube functioning

The basic geometry is considered to be critical for the protective function of the Eustachian tube - middle ear system. Reflux of liquid into the body of the flask occurs if the neck of the flask is excessively wide, or the length of the neck of the flask is too short as seen in children. Because infants have a shorter Eustachian tube than adults, reflux is more likely to occur in the baby. The position of the flask in relation to the liquid is another important factor. In humans, the supine position enhances flow of liquid into the middle ear; thus infants might be at risk for developing reflux otitis media because they are commonly supine. Reflux of liquid into the vessel can also occur if a hole is made in the bulbous portion of the flask, because this prevents the creation of positive pressure in the bulbous portion. This positive pressure is useful in the prevention of reflux of material from the neck of the flask.

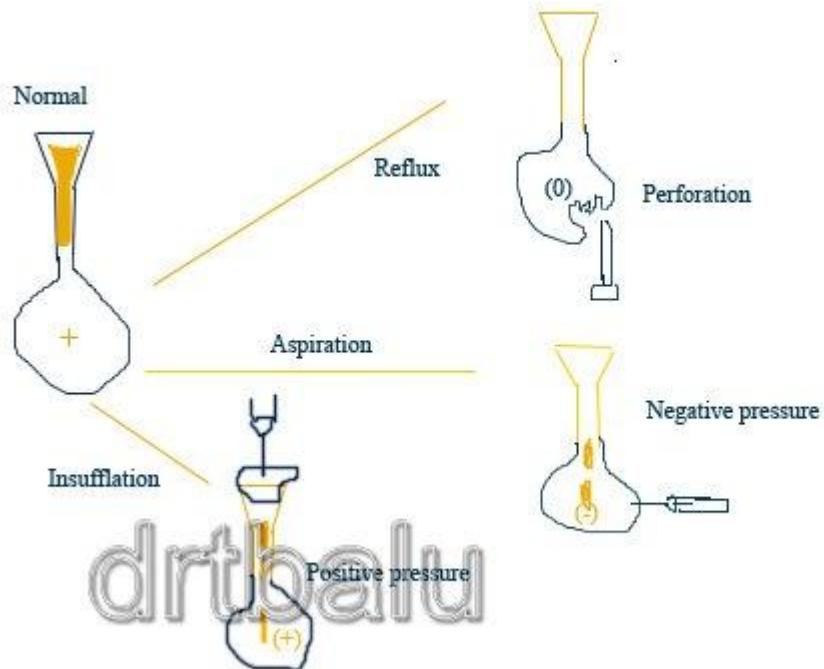


Figure showing pathogenesis of middle ear disease

If negative pressure is applied to the bulbous portion of the flask then this pressure is sufficient to cause aspiration of contents from the neck of the flask. This scenario is represented by high negative pressure in middle ear as it occurs in nose blowing, crying, closed nose swallowing, diving or airplane descent. The neck of the Eustachian tube is supposed to be compliant hence compliance plays a vital role in prevention of reflux of secretions.

Clinical features:

Acute suppurative otitis media passes through 4 stages: 1. Stage of hyperemia

2. Stage of exudation

3. Stage of suppuration

4. Stage of resolution.

The progression of these stages depends on the virulence of the infecting organisms, resistance of the host, adequacy of antibiotic therapy. If the infecting organism is virulent or if the antibiotic treatment is not sufficient then the disease may progress to a stage of coalescent mastoiditis with its attendant complications.

Stage of hyperemia: Initial infection by infection results in hyperemia of the mucous membrane causing otalgia, fever and fullness in the affected ear. This stage is characterized by oedema of the mucoperiosteum due to vascular engorgement. Otoscopy show dilated vessels along the handle of malleus and along the rim of the tympanic membrane. Antibiotic therapy during this stage will help in resolution of the disease. Amoxycillin is the drug of choice.

Stage of exudation: Absence of treatment during the stage of hyperemia leads to the stage of exudation. In this stage there is outpouring of fluid from the dilated vessels of the mucoperiosteum. This fluid is serous in nature containing fibrin, red cells, and polymorphs. This exudate fills the tympanomastoid compartment really fast, and the whole middle ear cavity is under intense pressure due to this retained secretion. Pain is the most prominent feature of this stage. The patients may have fever and fullness in the ear. Otoscopy shows a bulging ear drum with loss of all landmarks. The drum is reddish and bulging in nature. These patients have also coexistent mastoid tenderness due to mastoiditis.

Stage of suppuration: Failure of treatment during the stage of exudation leads on to stage of suppuration. The exudate present in the middle ear cavity is a very good culture medium and hence there is secondary bacterial infection leading on to suppuration.

Stage of resolution: is preceded by either rupture of the ear drum leading on to a serous / serosanguinous / purulent discharge from the ear. When the middle ear is free from the exudate / pus the stage of resolution sets in. The patient has reduction in otalgia, fever subsides. The patient has considerable clinical improvement.

Stage of complication: If the infection persists beyond a period of 2 weeks then there is associated thickening of the mucoperiosteum especially in the air cells around the peri antral area leading to a block in the drainage from the antral cells. The pent up secretions in the mastoid air cell system causes intense pressure, venous stasis and local acidosis. This acidosis causes dissolution of calcium from the bone causing decalcification and coalescence of the mastoid air cell system. This condition is known as coalescent mastoiditis. This stage is characterised by emergence of otalgia and low grade fever. Erosion of the outer cortex in the mastoid lead to the formation of abscess under the periosteum of the mastoid cortex. This condition is known as subperiosteal abscess.



Figure showing Otoscopic findings in a patient with acute otitis media

Management:

Acute suppurative otitis media is a self-limiting condition. If appropriate antibiotics are started early then it resolves. Amoxycillin is the drug of choice. Cephalosporins may also be started in refractive cases. Anti-inflammatory drugs like ibuprofen are also prescribed in order to alleviate pain. Patients who are refractory to medical management may undergo Myringotomy in order to decompress the middle ear cavity. This procedure is done using a myringotome.

Coalescent otitis media and subperiosteal abscess are surgical complications. These patients must be taken up for surgery under adequate antibiotic cover.

Citation: 1.

Acute otitis media by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/asom.html>

Eustachian tube ¹

Eustachian tube is otherwise known as Pharyngotympanic tube, middle ear ventilation tube. It is bony cartilaginous in nature. It connects the middle ear with the nasopharynx. In adults it lies at an angle of 45 degrees to the horizontal plane. In infants this inclination is about 10 degrees. In adults its length is 38mm. For descriptive purposes it can be divided into posterior 1/3 which is osseous in nature and anterior 2/3 which is cartilaginous in nature.

This Eustachian tube is shorter straighter and wider in infants predisposing middle ear infections through this tube. The osseous portion of the Eustachian tube also known as protympanum lies completely within the petrous portion of the temporal bone. The lumen of the osseous portion of the Eustachian tube is triangular and is open always in contrast to the fibrocartilaginous portion which is kept closed at rest, and opens during swallowing, or during a Valsalva maneuver. The osseous and cartilaginous portion of the Eustachian tube meets at an irregular bony portion and forms an angle of about 160 degrees with each other. The cartilaginous tube courses anteromedially and inferiorly, angled between 30 and 40 degrees. The cartilaginous portion of the tube is not completely surrounded by cartilage, but is deficient inferolaterally where it is covered by a membrane. The cartilage is crook shaped covering the medial, lateral and superior walls of the cartilaginous portion of the tube. The tubal lumen is shaped like two cones joined at their apices. The junction of the cones is the narrowest portion of the lumen and is known as the isthmus, and is usually situated at the junction of the cartilaginous and bony portion of the tube.

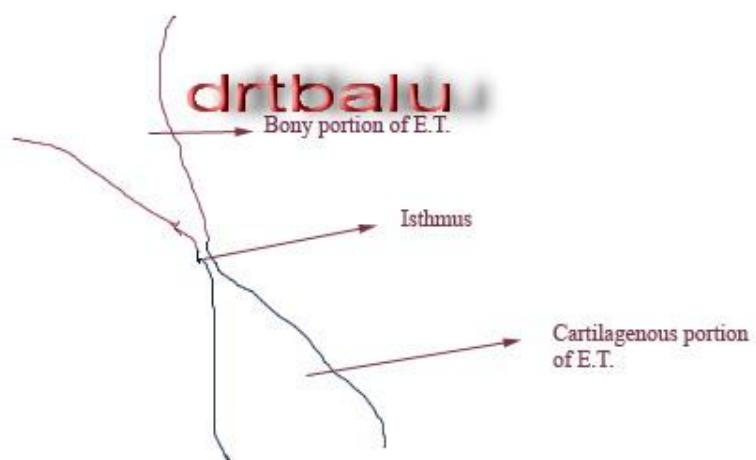


Diagram showing the Eustachian tube

The cartilaginous portion of the Eustachian tube does not follow a straight course in the adult but extends along a curve from the junction of the osseous and cartilaginous portions to the medial pterygoid plate, approximating the skull base during most of its course. The Eustachian tube crosses the superior border of the superior constrictor muscle to enter the nasopharynx. The medial cartilaginous portion of the tube presses against the pharyngeal wall to form a prominent fold, the torus tubaris. The torus is the site of origin of the salpingopalatine muscle and is the point of origin of the salpingopharyngeal muscle.

The mucosal lining of the Eustachian tube is continuous with that of the nasopharynx and middle ear (ciliated columnar epithelium). Certain differences in the mucosal lining is evident, mucous glands predominate at the nasopharyngeal orifice, and this gradually changes into a mixture of goblet cells at the tympanum.

Muscles associated with Eustachian tube: The muscles associated with the Eustachian tube are 4 in number. They are tensor veli palatini, levator veli palatini, salpingopharyngeus, and tensor tympani.

Usually the Eustachian tube is closed; it opens during such actions like swallowing, yawning thus equalising the middle ear pressure. Active dilatation of the tube is induced by the tensor veli palatini muscle. Closure of the tube has been attributed to passive reapproximation of tubal walls by extrinsic forces exerted by surrounding elastic fibres.

Blood supply: The Eustachian tube is supplied by the ascending palatine artery, pharyngeal branch of internal maxillary artery, the artery of the pterygoid canal, ascending pharyngeal artery, and the middle meningeal artery. The venous drainage is via the pterygoid plexus.

Nerve supply: The pharyngeal orifice of the Eustachian tube is supplied by a branch from the otic ganglion, the sphenopalatine nerve, and the pharyngeal plexus. The remainder of the tube receives its sensory supply from the tympanic plexus and the pharyngeal plexus. The glossopharyngeal nerve has an important role in the innervation of the Eustachian tube.

Functions of the Eustachian tube:

Ventilation: It ventilates the middle ear cavity via the nasopharyngeal airway.

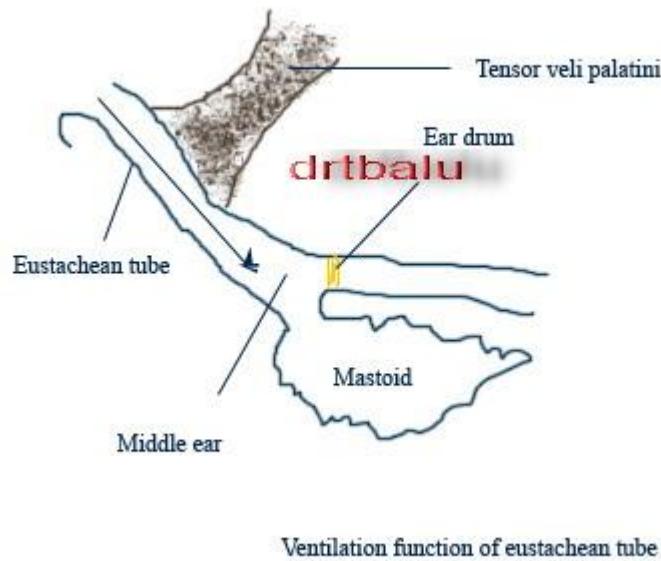


Figure showing ventilatory functions of middle ear

Protection: It protects the middle ear cavity from microbes of nasopharynx.

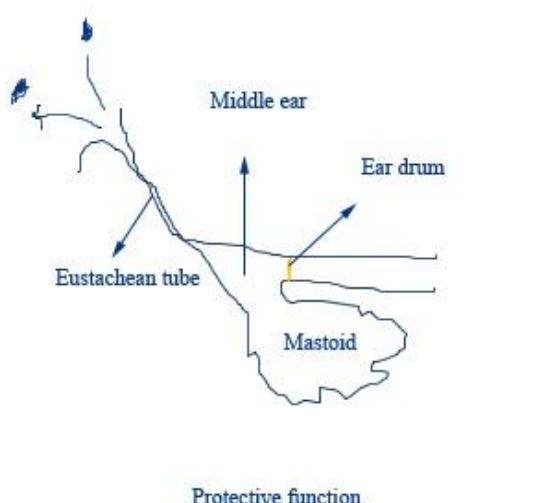
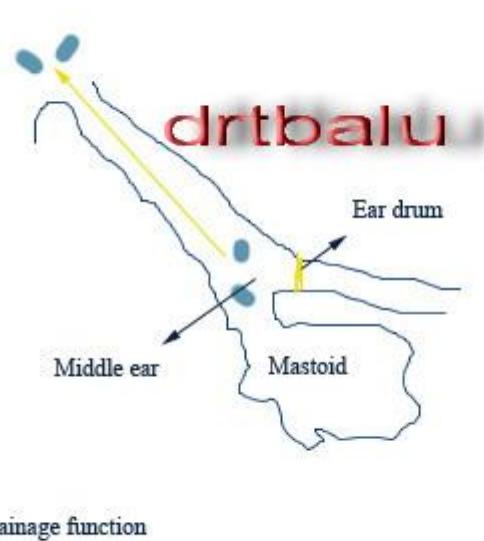


Figure showing protective function of Eustachian tube

Drainage: Drains the secretions from the middle ear cavity into the nasopharynx.



Drainage function

Figure showing the drainage functions of Eustachian tube

Features of Infant Eustachian tube: In infants the Eustachian tube is about half as long as in the adults, averaging about 18 mm. The osseous portion is longer than the cartilaginous portion. It is shorter, straighter, and wider than that of adults. The tensor veli palatini muscle is less efficient in infants. The tube is also mostly horizontal in infants. Hence infants are more prone for middle ear infections arising from the Eustachian tube.

Citation:

- 1.

Eustachian tube by drtblu [Internet]. [cited 2011 Dec 31]; Available from: <http://drtblu.co.in/et.html>

Tympanosclerosis ¹

Definition: is deposition of acellular hyaline and calcium deposits within the submucous layer of the tympanic membrane / submucous membrane of the middle ear cavity. In a majority of patients these plaques are so insignificant that they cause very little impairment in the hearing ability of the patient. These plaques could be likened to healed scar tissue.

When present over the tympanic membrane these plaques appear like a whitish crescentic shaped plaques.

Causes:

1. Could be caused by resolved otitis media.
2. Trauma
3. Chronic otitis media with effusion
4. After recurrent bouts of acute otitis media (middle ear tympanosclerosis)
5. After grommet insertion
6. Eustachian tube obstruction
7. Autoimmune process occurring within tympanic membrane

Histology:

There is hyalinisation of the subepithelial connective tissue of the tympanic membrane and middle ear cavity. Calcification is commonly present in these lesions. Osteoneogenesis can also occur within these lesions.

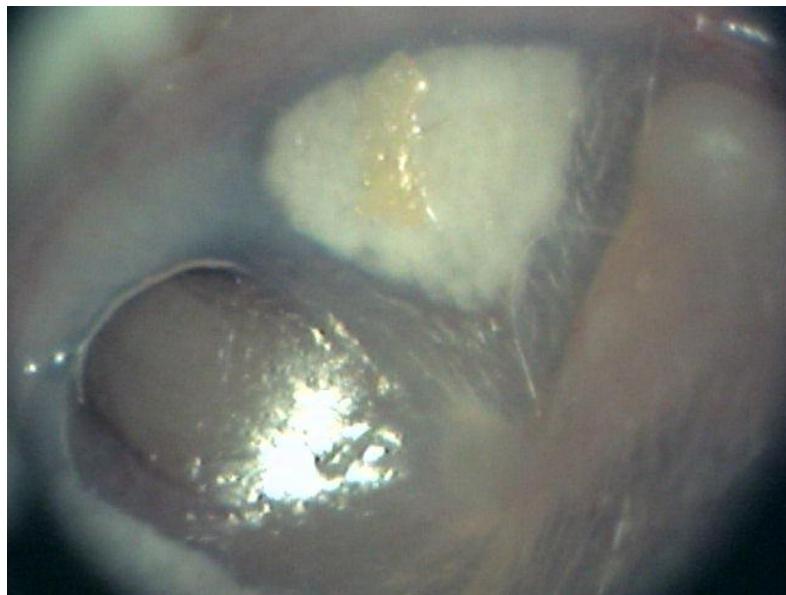
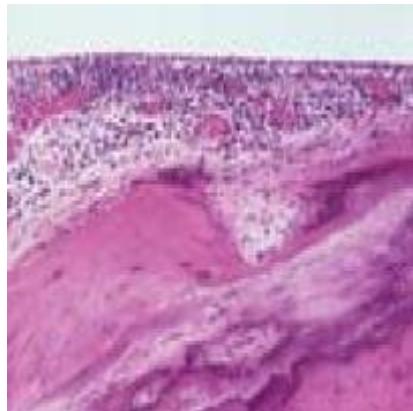


Figure showing tympanosclerotic plaque



Histopathology of tympanosclerotic plaque

Plaques occurring in the tympanic membrane are limited to lamina propria. Deposition of bone due to Osteoneogenesis in the attic region may cause fixation of malleus and incus leading on to conductive deafness.

Pathophysiology:

It has been postulated that after an episode of otitis media with effusion / or acute otitis media the collagen undergoes degeneration and subsequent dystrophic calcification and formation of tympanosclerosis.

Treatment:

Most of the patients with tympanosclerosis are symptom free and the finding is purely accidental. If these patients have significant conductive deafness then surgical removal of the plaques from the tympanic membrane and fashioning a neo tympanic membrane using temporalis fascia graft can be attempted. If these plaques involve the attic area and cause ossicular fixation leading on to conductive deafness, ossiculoplasty can be attempted.

Citation:

1.
Tympanosclerosis by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
http://drtbalu.co.in/tympano_scle.html

Tympanic Membrane ¹

Tympanic membrane is also known as the ear drum. Anatomically it could be considered to be a part of the external ear since it is attached to the medial terminal end of the bony meatus. Functionally speaking it is part of the tympanic cavity.

It is more or less oval in shape (egg shaped). It is 9mm in diameter. Its broad portion lies superiorly. It is pearly white in color, thin and semi-transparent. When viewed under illumination a triangular cone of light (reflected light) is seen extending from the centre forwards and downwards. This reflection or cone of light is due to the sectional shape of the membrane. The ear drum is set with an obliquity of about 55 degrees to the floor of the external meatus. The centre of the ear drum appears retracted, and is known as the umbo. This umbo lies at the apex of the cone of light. Visible as an ivory colored extension upwards from the umbo is the handle of the malleus. If the posterior portion of the membrane is transparent, then the image of the long process of the incus, and occasionally the stapedial tendon may also be seen.

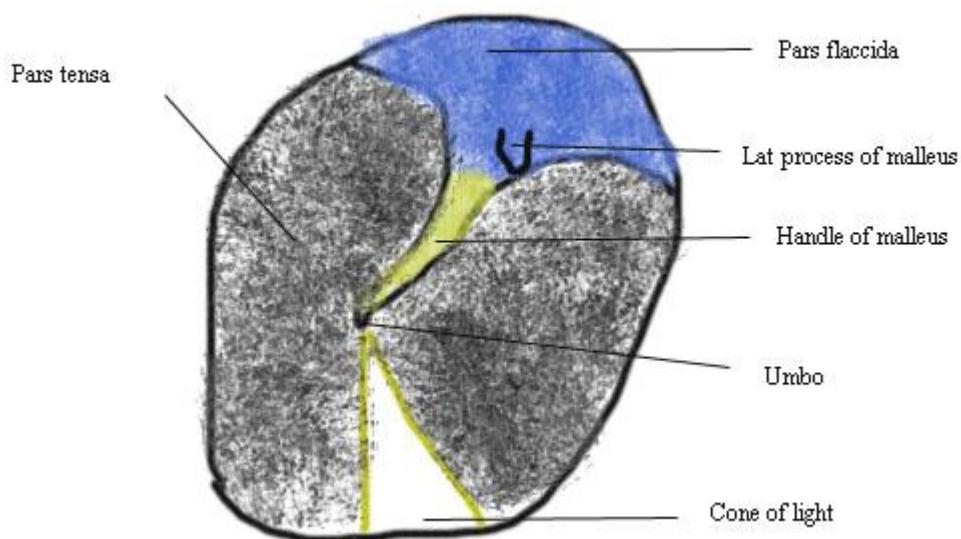


Figure showing the parts of ear drum

The ear drum is composed of 3 layers. The outer layer is formed by stratified squamous epithelium, and is continuous with that of the external auditory canal. Any condition affecting the skin of the external canal will also affect the outer layer of the ear drum. Common conditions like dermatitis involving the skin of the external canal can also involve the outer layer of the ear drum. Embryologically outer layer of the ear drum developed from the ectoderm. Myringitis granulosa a common condition affecting the ear drum affects only

the outer layer of the tympanic membrane. The middle and inner layers are not involved in this condition. It is commonly caused by infections arising from the external canal. Constant irritation of the ear drum due to presence of wax may also predispose to this condition. Another condition which involves the outer layer of the ear drum is Bullous myringitis. In this condition blebs may be seen in the outer layer of the ear drum. It is commonly caused by viral infections, or mycoplasma pneumonia. It may also be associated with middle ear effusion.

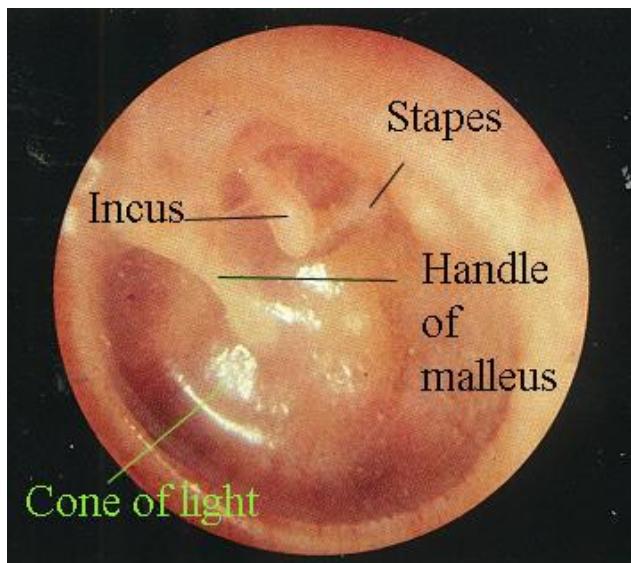


Figure showing normal ear drum



Figure showing Myringitis granulosa

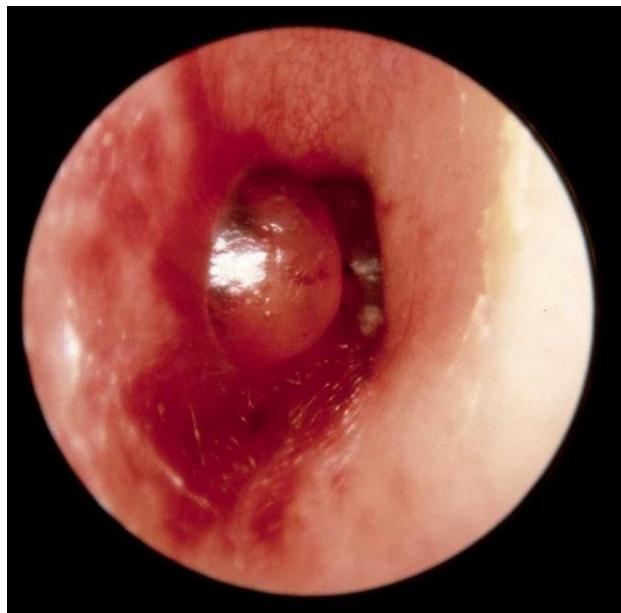
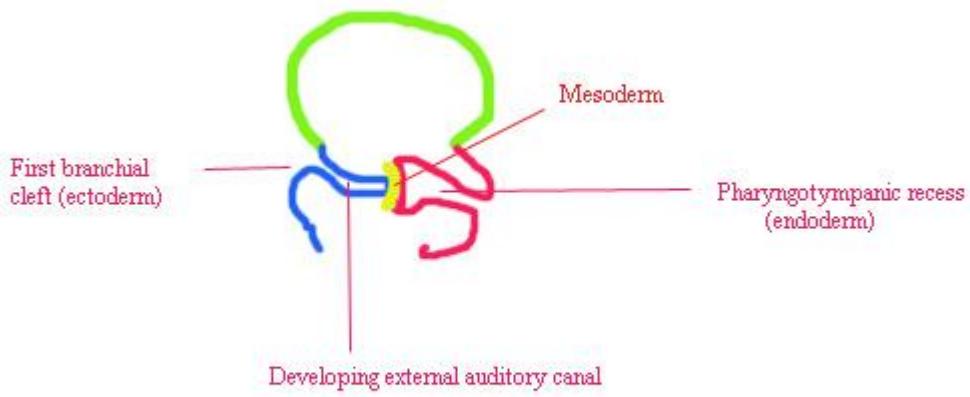


Figure showing acute otitis media

The middle fibrous layer from which the ear drum derives its strength and resilience is derived from the mesoderm. This portion is in fact sandwiched between the outer squamous lining derived from the ectoderm and inner mucosal lining of the middle ear cavity derived from the endoderm. The ectodermal and mesodermal components of the ear drum arise from the first branchial cleft, while the endodermal component is derived from the pharyngotympanic recess. The middle fibrous layer has two components: 1. radial and 2 circular fibres. The handle of the malleus lie between the middle fibrous layer and the inner mucosal layer of the ear drum. From the handle of the malleus the radial fibres of the middle fibrous layer radiate towards the circumference of the ear drum. The circular fibres are more prominent and thickened along the circumference of the ear drum. The condensations of the circular fibres are fixed to the tympanic sulcus at the medial end of the external auditory canal. This middle fibrous layer is absent in the attic area of the ear drum. The fibrocartilaginous ring and the fibrous layer of the ear drum are deficient superiorly. This deficient area is known as the notch of Rivinus. The attic portion of the ear drum which lacks the middle layer is known as the pars flaccida, while the rest of the drum which has all the three layers is known as pars tensa. The chorda tympani nerve which is a branch of the facial nerve run between the middle fibrous and inner mucosal layers of the ear drum.



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Diagram showing embryology of ear drum

The skin of the external canal and the outer lining of the tympanic membrane are unique in a sense that they lack frictional and abrasive contacts which is common with the skin lining elsewhere in the body. Desquamated keratin does not accumulate on the surface of the tympanic membrane, or in the deep external meatus, because the skin lining here is endowed with a peculiar feature known as Migration. The surface layers of the skin of the ear drum, and the surface keratin move towards the periphery of the membrane, and then slowly along the external meatus to the exterior. Derangements of this unique feature are associated with some of the diseases of the external ear.

The inner layer of the ear drum derived from the endoderm of the pharyngotympanic recess is continuous with that of the mucosal lining of the middle ear cavity.

Blood supply:

The external surface of the ear drum receives its blood supply from the deep auricular branch of the maxillary artery. This small artery leaves the first part of the maxillary artery behind the neck of the mandible and gains access into the external canal by piercing the anterior wall behind the mandibular joint. It sends small branches into the membrane from the whole circumference of the pars tensa and one or more manubrial branches that descend on the handle of mandible from above. The internal surface of the ear drum is supplied from behind by the stylomastoid branch of the posterior auricular artery, and from

the front by the tympanic branch of the maxillary artery. The superficial veins open into the external jugular vein; and those on the internal surface drain into the transverse sinus and veins of the dura mater, and partly into the venous plexus on the Eustachian tube.

Nerve supply:

The innervation of the posterior half of the ear drum is by the auricular branch of the X nerve and the anterior half is by the auriculotemporal branch of the Vth nerve. The inner surface of the ear drum is supplied by the tympanic branch of the IXth nerve.

Appearance of the ear drum in various diseases:

The normal ear drum is pearly white in color. In pathological states this color of the drum may change.

Red drum: Is seen in acute otitis media and in glomus jugulare. In acute suppurative otitis media discharge may be seen extruding from a small perforation in the pars tensa portion of the ear drum. This is known as the light house sign.

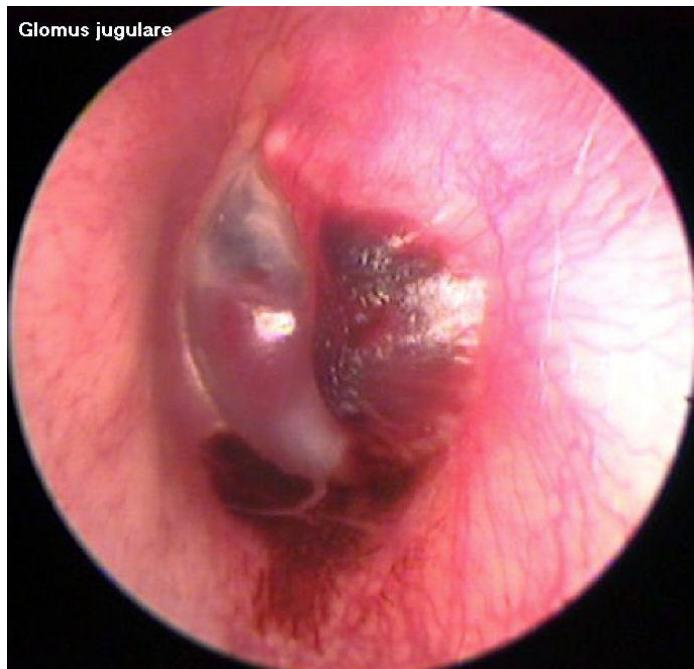


Figure showing features of AOM (Red drum)

Blue drum: is commonly seen in secretory otitis media, high jugular bulb etc.

Blue drum as seen in secretory otitis media

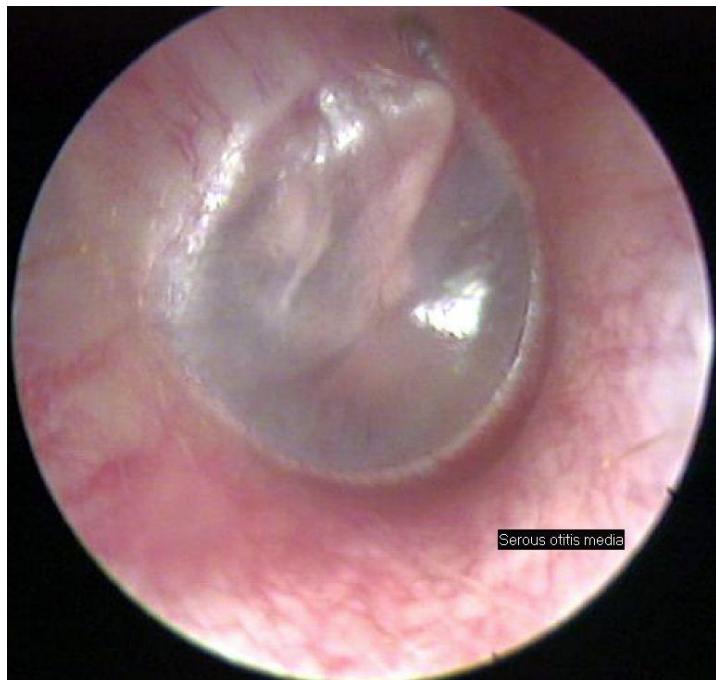


Figure showing ear drum appearance in serous otitis media

Citation:

1.
Tympanic membrane by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/tm.html>

Impedance audiometry ¹

Synonyms: Tympanometry, acoustic immittance test.

The primary purpose of impedance audiometry is to determine the status of the tympanic membrane and the middle ear. It is also otherwise known as Tympanometry or acoustic immittance test. The secondary purpose of this investigation is to evaluate the acoustic reflex pathway which includes the 7th and 8th cranial nerves and the brain stem. This test should not be used to assess the sensitivity of hearing and the results of this test should always be viewed in conjunction with the results of pure tone audiogram.

Impedance audiometry is a measurement of energy or air pressure which involves the external auditory canal, the ear drum, ossicular chain, stapedius muscle, cochlea, 7th cranial nerve, 8th cranial nerve and the brain stem. This test is affected by the mass, mobility and resistance systems of the external and middle ear cavities.

The following tests have been included under the battery of impedance audiometry:

1. Tympanometry
2. Eustachian tube function
3. Tests to identify perilymph fistula
4. Acoustic reflex threshold
5. Acoustic reflex decay

These tests can be used to identify the following pathologies involving the peripheral and central portions of hearing.

- a. Middle ear effusion
- b. Ear drum perforations including patency of Eustachian tube
- c. Tympanosclerosis
- d. Hypermobile ear drum
- e. Eustachian tube dysfunction
- f. Glue ear
- g. Otosclerosis

h. Ossicular discontinuity

g. Acoustic neuroma

h. Facial nerve function

i. Hearing loss

j. Brain stem disorders

Tympanometry: Measures the sound reflected from the ear drum while the pressure of the external canal is varied by the operator. It aids in the assessment of outer ear, middle ear and the Eustachian tube. This test should not be performed in infants below the age of 7 months because the suppleness of the cartilage of the external canal may produce misleading results.

Procedure: First the probe is inserted into the external auditory canal till a airtight seal is obtained. Probe tone is presented typically at 226Hz into the ear canal while the air pressure of the canal is altered between +200 and - 400 decapascals. The maximum compliance occurs when the pressure of the external auditory canal and the middle ear becomes equal. Only at this pressure maximal acoustic transmission occur through the middle ear. The compliance peak therefore indicates therefore indicates the pressure of the middle ear implying efficacy of the Eustachian tube function. The height of the compliance peak indicates the mobility / stiffness of the tympanic membrane or the middle ear cavity.

The term static compliance indicates the height of the tympanogram at its peak, and it is the measurement of the mobility of the whole system.

Classification of tympanograms:

The classification system introduced by Jerger is commonly used to classify various types of tympanograms. Other systems have been proposed, but none of them are in common use.

Type A curve: Suggests normal middle ear function. The compliance peak occurs between - 150 - +100 dapa. The value of compliance ranging between 0.2 - 2.5 millimhos. This type of curve is also known to occur in early stages of otosclerosis



Figure showing Impedance audiometer

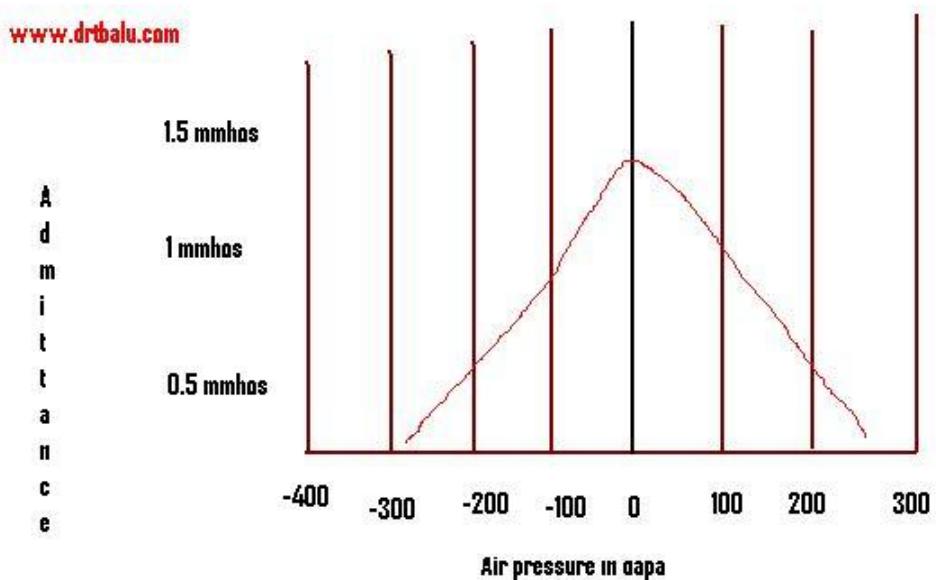


Fig showing Jerger Type A impedance curve.

Type As curve: is a shallow curve suggesting a stiffened middle ear system. Compliance peak occurs at -150 - + 100 dapa. The compliance value is less than 0.2 mmhos. This curve is commonly found in patients with glue ear, stiffened ear drum, or otosclerosis.

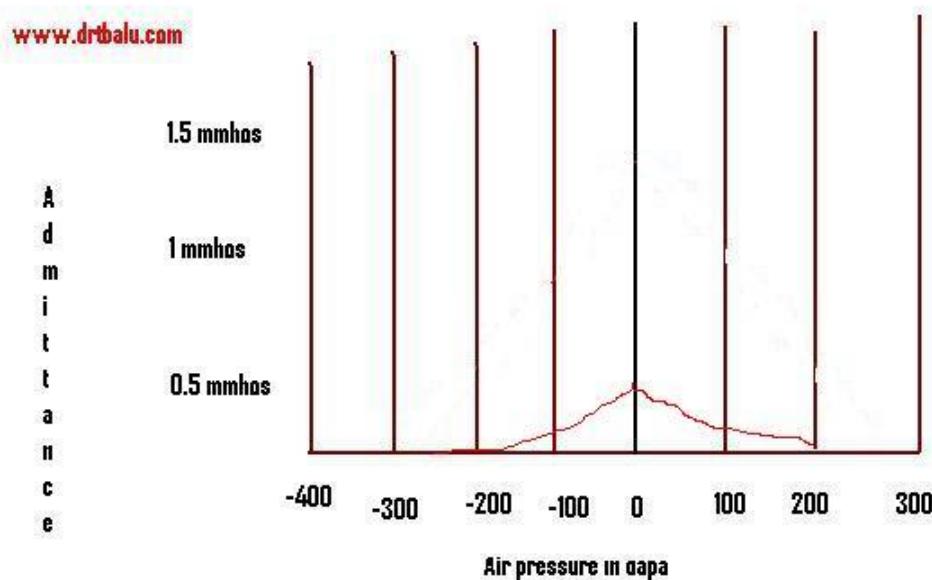


Fig showing Jerger Type As curve

Type Ad curve: is a deep curve suggests a flaccid ear drum or middle ear system, ossicular disruption. Usually ossicular disruption gives compliance higher than the recording parameters (infact the recording goes off chart). The compliance peak occurs between -150 - + 100 dapa. The compliance value is more than 2.5 mmhos.

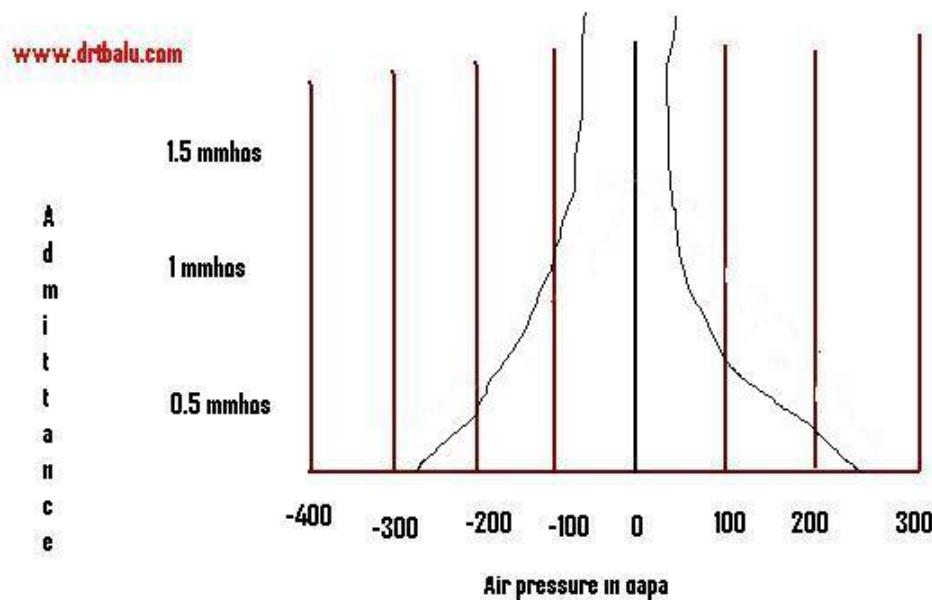


Fig showing Jerger Type Ad curve: Note the compliance value is so high that the curve goes off the chart.

Type B: is a flat curve with no compliance peak. This Type B curve must always be interpreted in conjunction with the ear canal volume. Average ear canal volume in children ranges between 0.42 - 0.97 ml, while in adults it ranges between 0.63 - 1.46 ml.

Type B curve with normal ear canal volume suggests otitis media.

Type B curve with small canal volume suggests that the ear canal could be occluded by the presence of wax, or the probe of the impedance audiometer has not been properly placed.

Type B curve with large canal volume suggests that there could be perforation of the ear drum. This curve is caused due to a patent pressure equalisation system

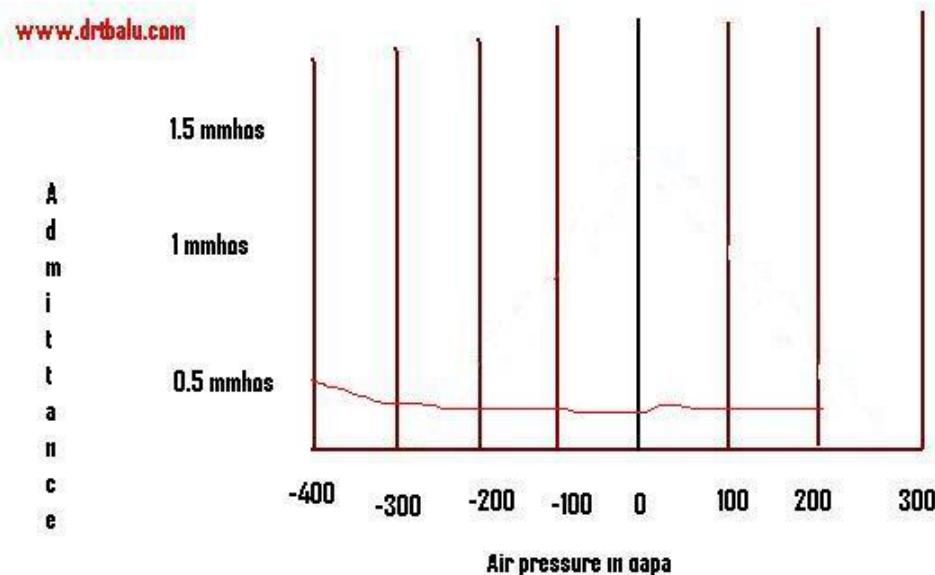


Fig showing Jerger Type B curve

Type C curve: This curve suggests a significant negative pressure in the middle ear, or Eustachian tube dysfunction. Compliance is recordable but the peak compliance occurs at less than -150 dapa.

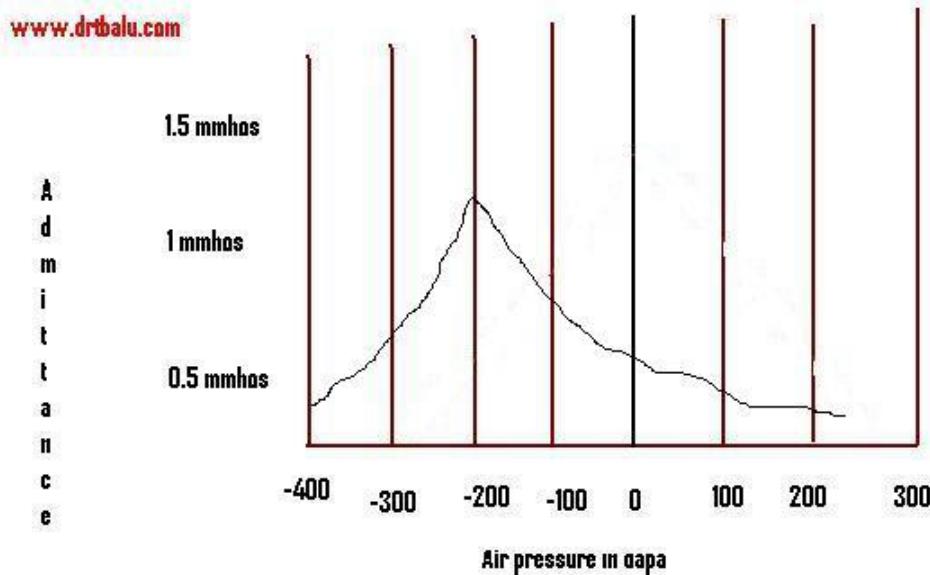


Fig showing Jerger Type C curve.

Assessing Eustachian tube functioning by impedance audiometry:

The function of the Eustachian tube can easily be assessed by reading the tympanograms. Type A tympanograms reflect a normal middle ear function which is only possible in the presence of a normally functioning Eustachian tube. Similarly Type C tympanograms indicate significant negative pressure in the middle ear implying that the Eustachian tube is blocked. If there is tympanic membrane perforation a Type B curve will be produced. In this situation the Eustachian tube function cannot be assessed using a tympanogram. However an indirect assessment of the pressure equalisation function of the Eustachian tube can be made by increasing the probe pressure in the external ear canal, asking the patient to swallow then assessing whether the Eustachian tube is able to clear the increased pressure applied to the external ear canal.

Testing for the presence of absence of perilymph fistula:

This can be indirectly assessed by the presence of intense giddiness along with nystagmus when the external canal pressure is increased by increasing the probe pressure. This sign is also known as the Hennebert's sign. This sign is manifested only in the presence of perilymph fistula.

Eliciting acoustic reflex thresholds: This is a measure of the stapedial muscle reaction to exposure to high intensity sounds. When the stapedius muscle contracts in response to sound it stiffens the ossicles and the ear drum altering the compliance values which can be

measured using an impedance audiometer. The recording is ideally made at a single pressure setting i.e. the pressure which shows the maximum compliance. The reflex on the opposite side also is tested since it is a bilateral reflex. The sound frequencies used to test this reflex are 500, 1000, 2000 and 4000Hz. For screening purposes it is sufficient if 1000Hz is used.

The acoustic reflex cannot be recorded in patients with a type B tympanogram. It also cannot be recorded in patients with severe profound sensorineural hearing loss. The reflex may be attenuated in the presence of conductive deafness. Using this test it is possible to assess the whole of the acoustic reflex pathway. If the pathway is affected at central level then ipsilateral recordings will be normal with absent contralateral acoustic reflexes.

Citation:

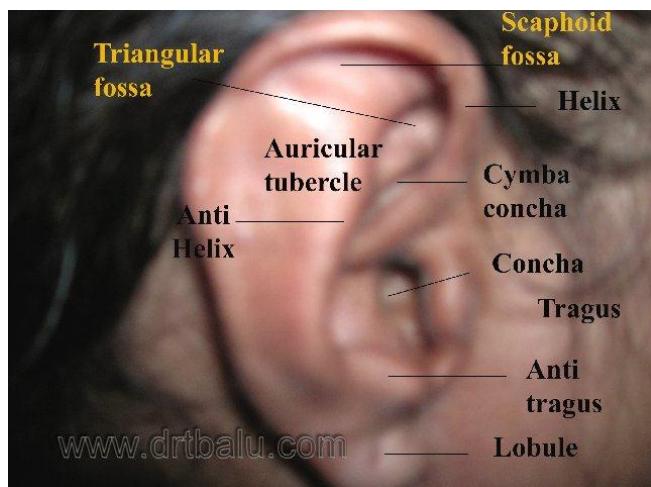
1.
Impedance audiometry by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
http://drtbalu.co.in/imp_audio.html

Pinna

Synonyms: Auricle, pinna

Development:

The auricle starts to develop when 6 hillocks appear around the first pharyngeal groove, which lies between the first and the second branchial arches. Three hillocks develop on each side of the groove; as their growth progresses they contribute to the development of the auricle. The bulk of the auricle is developed from the mesenchyme of the second branchial arch (Hyoid arch). The cartilage of the auricle extends inwards partially to surround the future external meatus. A rudimentary pinna is formed by the 60 day of embryonic life, and by 4 months the convolutions are formed resembling the adult pinna.



Photograph showing pinna

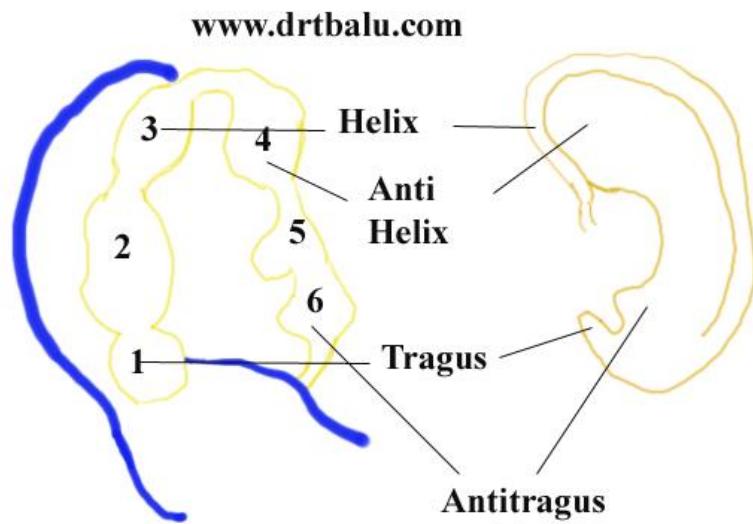


Figure showing embryology of pinna

Anatomy:

The pinna projects from the side of the skull to varying degrees. It has two surfaces; the lateral which is the exposed surface, and the medial which is hidden. The lateral surface has several prominences and depressions giving it a unique shape. The curved rim of the pinna is known as the helix. At the posterior superior aspect of the helix is a small tubercle known as the auricular tubercle (Darwin's tubercle). Anterior and parallel to the helix another prominence is present known as the antihelix. Superiorly the antihelix divides into two crura encompassing a fossa known as the triangular fossa. Above the superior of the two crura lies another fossa known as the scaphoid fossa. In front of the antihelix, in fact partly encircling it is the concha. The antero superior portion of the concha is covered by the descending limb of the antero superior portion of the helix. This region of the concha is known as the cymba concha. This cymba concha has an important surface relationship with the suprameatal triangle. In fact the suprameatal triangle or (McEwen's triangle) lie just under the cymba concha. This triangle acts as a surface marking for the mastoid antrum during mastoidectomy procedures.

Opposite to the concha, and lying across the external auditory meatus (partially covering it) is a small blunt triangular prominence known as the Tragus. This prominence points posteriorly. Opposite to the tragus lies another prominence known as the antitragus.

The body of the auricle is covered with fibroelastic cartilage, covered with skin. It is connected to the surrounding parts by ligaments and muscles. It is also continuous with the cartilage lining the external auditory canal. The skin of the auricle is thin and closely bound

to the perichondrium on the lateral side. On the medial side the skin is not adherent to the cartilage of the auricle, there is a layer of subdermal adipose tissue separating the skin from the cartilage. The skin is covered with fine hairs which have sebaceous glands opening into their root canals. The glands are most numerous in the concha and the scaphoid fossa. The fibroelastic cartilage is absent in the lobule area of the pinna. The auricular cartilage depends on the perichondrium for its nourishment. The cartilage is connected to the temporal bone by two extrinsic ligaments, the anterior and the posterior ligaments. Intrinsic ligaments connect various parts of the cartilaginous auricle.

The muscles of the auricle belong to two groups; the extrinsic and the intrinsic groups. The extrinsic muscles are supplied by temporal and post auricular branches of the facial nerve. The extrinsic muscles are functionally not so important, but they give rise to the post auricular myogenic response following auditory stimulation. The extrinsic muscles are auriculares anterior, auriculares superior and auricularis posterior.

The intrinsic muscles are 6 in number, all small and inconsistent and without useful function.

Blood supply:

The post auricular branch of the external carotid artery supplies the medial surface of the pinna, and extends around the helix to supply the extremities of the lateral surface. The anterior auricular branch of the superficial temporal artery supplies the bulk of the lateral surface. Auricular branch from the occipital artery assists the post auricular artery in supplying the medial surface.

Lymphatic drainage:

Lymphatic drainage from the posterior surface of the pinna is to the lymph nodes at the mastoid tip, from the tragus and from the upper part of the anterior surface is to the preauricular nodes, and from the rest of the auricle to the upper deep cervical nodes.

Sensory innervation of the auricle:

Nerve	Derived from	Region Supplied
Greater Auricular	Cervical Plexus C2 C3	Medial surface and posterior part of lateral surface of pinna
Lesser occipital	Cervical plexus C2	Superior portion of medial surface
Auricular	Vagus	Concha and Antihelix
Auriculo temporal	Mandibular division of trigeminal nerve	Tragus, Crus of the helix and adjacent helix
Facial		Supplies root of concha

Physiology of pinna:

1. Pinna increases the pressure at the tympanic membrane in a frequency sensitive way, thus emphasizing certain frequencies in the input.
2. It helps to localise the direction of the source of sound.
3. Protects the middle ear and inner ear from extraneous insults

Citation:

1.
Pinna by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/pinna.html>

Anatomy of Middle ear

Synonyms: Middle ear cleft, Tympanum

The middle ear cleft includes the tympanum (middle ear cavity proper), the Eustachian tube, and the mastoid air cell system. The tympanic cavity is an air filled irregular space contained within the temporal bone. It also contains the three auditory ossicles (malleus, incus and stapes) along with their attached muscles. For the purpose of description the tympanic cavity may be considered as a box with four walls, a roof and a floor. The corners of this hypothetical box are not sharp.

Lateral wall: The lateral wall of the tympanum / middle ear is partly bony and partly membranous. The central portion of the lateral wall is formed by the tympanic membrane, while above and below the tympanic membrane there is bone, forming the outer lateral walls of the epitympanum (attic) and hypotympanum respectively. The lateral wall of the epitympanum (attic) also includes that part of the tympanic membrane lying above the anterior and posterior malleolar folds - this portion of the ear drum is also known as pars flaccida. This portion of the tympanic membrane lacks the middle fibrous layer, hence the name. The lateral attic wall (bony portion) is wedge shaped; its lower portion is also called the outer attic wall (scutum). Scutum actually means shield in Latin. This bony portion is thin and its lateral surface forms the superior portion of the deep portion of the external meatus.

Three openings are present in the bone of the medial surface of the lateral wall of the tympanic cavity. The first opening is the posterior canaliculus for the chorda tympani nerve. This opening is situated at the junction between the lateral and posterior walls of the tympanic cavity. This opening is usually present at the level of the upper end of the handle of the malleus. This opening leads to the bony canal which descends through the posterior wall of the tympanic cavity. Since chorda tympani nerve traverses this canal it is also known as the canal for chorda tympani nerve. This canal also contains a branch from the stylomastoid artery which usually accompanies the chorda tympani nerve.

The second opening is the petrotympanic (Glaserian) fissure. This fissure opens anteriorly just above the attachment of the tympanic membrane. This opening is in fact a small slit about 2 mm long. It receives the anterior malleolar ligament. It also transmits the anterior tympanic branch of the maxillary artery to the tympani cavity.

The third is the canal of Hugier. It lies medial to the Glaserian fissure. The chorda tympani nerve enters through this.

Roof: The roof of the middle ear cavity is formed by the tegmen tympani. It is this tegmen tympani which separates the middle ear cavity from the dura of the middle cranial fossa. This tegmen tympani is formed in part by the petrous portion of the temporal bone, and the squamous portion of the temporal bone. The suture line between these two components is

known as the petrosquamous suture line. This suture line is unossified in the young, and does not close until adult life is reached. Through this suture veins from the middle ear may pass to the superior petrosal sinus.

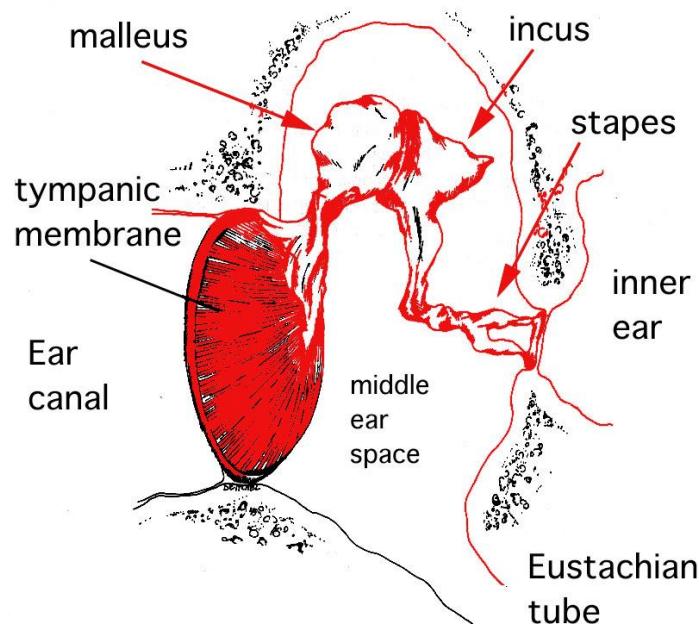
Floor: The floor is much narrow. In fact it is narrower than the roof of the middle ear cavity. This portion of the middle ear cavity lies in close relationship with the jugular bulb. The middle ear cavity is separated from the jugular bulb by a thin piece of bone. Rarely, the floor may be deficient and the jugular bulb in these patients is separated from the middle ear cavity only by fibrous tissue and mucous membrane. At the junction of the floor and the medial wall of the middle ear there is a small opening which allows the entry of tympanic branch of glossopharyngeal nerve to pass into the middle ear. This nerve takes an important part in the formation of tympanic plexus.

Anterior wall: The anterior wall of the tympanic cavity is very narrow. This is because the medial and lateral walls converge anteriorly. The anterior wall can be divided into two portions; the upper and lower portions. The lower portion of the anterior wall is larger than the upper portion. It has a thin plate of bone which separates this portion from the internal carotid artery as it enters the skull. This plate has two openings for the carotico tympanic nerves. The upper opening transmits the superior carotico tympanic nerve and the inferior opening transmits the inferior carotico tympanic nerve. It is through these nerves that sympathetic nerves reach the tympanic plexus. The upper smaller part of the anterior wall has two tunnels placed one below the other. The upper tunnel transmits the tensor tympani muscle, and the lower tunnel transmits the bony portion of the Eustachian tube.

Medial wall: The medial wall separates the middle ear from the inner ear. The most prominent portion of the medial wall of the middle ear cavity is the promontory. It is a rounded projection occupying most of the central portion of the medial wall of the middle ear. This projection is raised by the underlying basal turn of the cochlea. The promontory has numerous small grooves on its surface. These grooves contain the tympanic plexus of nerves. Behind and above the promontory is the oval window (fenestra vestibuli). This is a oval shaped opening connecting the tympanic cavity with the vestibule. In life this is closed by the foot plate of stapes and its surrounding annular ligament. The long axis of the fenestra vestibuli is horizontal. Its inferior border is concave. The size of the oval window varies, but on an average it is 3.25mm long and 1.75 mm wide. Above this fenestra vestibuli is the canal for facial nerver (horizontal portion) and below lies the promontory. Hence the fenestra vestibuli lies at the bottom of a depression also known as fossula that can be of varying depths depending on the position of the facial nerve and the prominence of the promontory.

The fenestra cochlea (round window) lies just below and behind the oval window. It is closed in life by a membrane known as the round window membrane (secondary tympanic membrane). The secondary tympanic membrane appears to be divided into an anterior and posterior portions by the presence of a transverse thickening. The diameter of the round window membrane is between 1.8 to 2.3 mm. It is made up of three layers; the outer mucosal, middle fibrous and an inner endothelial layer. The membrane of the fenestra cochleae does not lie at the end of the scala tympani but forms part of its floor. The ampulla of the posterior semicircular canal is the closest vestibular structure to this membrane. The nerve supplying the ampulla of the posterior semicircular canal (singular nerve) lies close to

this secondary tympanic membrane. The secondary tympanic membrane forms a landmark for the position of the singular nerve. This is useful during surgical procedures like singular neurectomy for treatment of intractable vertigo. These two windows (oval & round) are separated by the posterior extension of the promontory. This is known as the subiculum. Rarely a spicule of bone arises from the promontory above the subiculum and runs to the pyramid on the posterior wall of the middle ear cavity. This spicule of bone is known as the ponticulus. The round window faces inferiorly and a little posteriorly, lying completely under the cover of the promontory and hence usually is difficult to visualise. The round window niche is usually triangular in shape, having anterior, posterosuperior and posteroinferior walls. The posterosuperior and posteroinferior walls meet posteriorly leading on to the sinus tympani. This sinus tympani is a difficult area to visualise. Cholesteatoma may lurk in this area making it difficult to remove. This is one of the commonest causes of cholesteatoma recurrence after mastoidectomy. Small mirrors known as the zinne mirror can be used to visualise this area indirectly. Since sinus tympani lies under the pyramid, removal of the pyramid during surgery will bring the sinus tympani area into view. The facial nerve canal is another important anatomical structure present in this wall. This nerve runs above the promontory and fenestra vestibuli in an anteroposterior direction. The canal may occasionally be deficient leaving an exposed facial nerve. This is a dangerous anatomical variant because this nerve can easily be traumatised during any surgical procedures in the middle ear cavity. Even infections of the middle ear mucosa can cause facial nerve palsy in patients with an exposed facial nerve. The anterior end of the facial nerve canal is marked by the presence of a bony process known as processus cochleariformis. This curved projection of bone is concave anteriorly and it houses the tendon of the tensor tympani muscle as it turns laterally to the handle of the malleus. Behind the fenestra vestibuli, the facial nerve turns inferiorly to begin its descent in the posterior wall of the tympani cavity.



Diagrammatic illustration of middle ear anatomy

The region above the level of the facial nerve canal forms the medial wall of the epitympanum or attic. The dome of the lateral semicircular canal extends a little lateral to the facial canal and is the major feature of the posterior portion of the epitympanum. In well pneumatized bones this dome of the lateral canal can be very prominent.

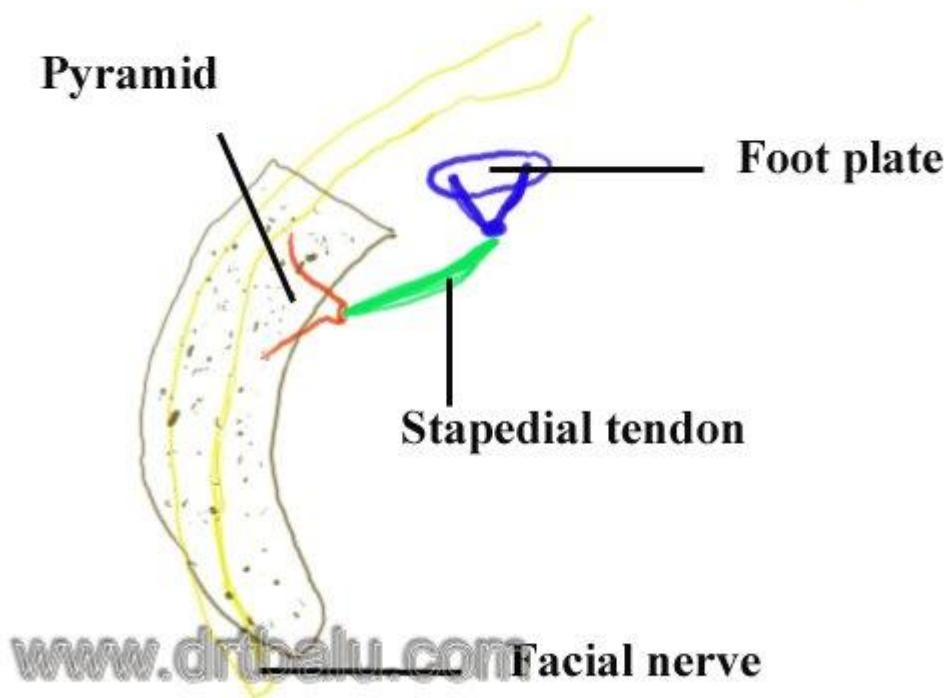


Figure showing pyramid of middle ear and its relations

Posterior wall: The posterior wall of the middle ear is wider above than below. In its upper part it has an important opening known as the aditus. This aditus helps the middle ear communicate with the mastoid air cell system. Aditus is a large irregular opening connecting the mastoid antrum to the middle ear cavity. Below the aditus is a small depression known as the fossa incudis. Fossa incudis houses the short process of the incus. Below the fossa incudis lies the pyramid.

Pyramid is a small conical projection which is hollow and its apex pointing anteriorly. It contains the stapedius muscle, the tendon of which passes forwards to insert into the neck of the stapes. The canal within the promontory curves downwards and backwards to join the descending portion of the facial nerve canal. Between the promontory and the tympanic annulus is the facial recess. The facial recess is bounded medially by the facial nerve and laterally by the tympanic annulus. Running through the wall between the two with varying degrees of obliquity is the chorda tympani nerve. This nerve always runs medial to the tympanic membrane. Drilling over the facial recess area between the facial nerve and the annulus in the angle formed by the chorda tympani nerve can lead into the middle ear cavity. This surgical approach to the middle ear cavity through this area is known as the

facial recess approach. This approach is suitable for surgeries involving the round window niche like placement of electrodes during cochlear implant procedures. Hypotympanum can also be approached through this approach.

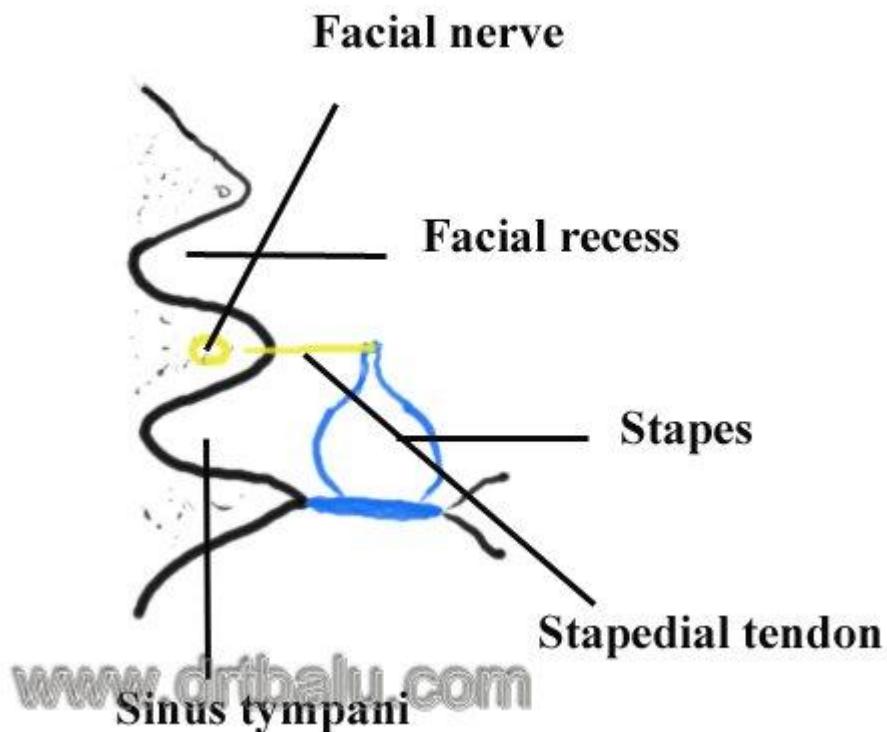


Figure showing sinus tympani of middle ear

Contents of the middle ear:

The most important content of the middle ear is air. The air flows into the middle ear through a patent Eustachian tube. The other contents are:

Chain of three ossicles which help in sound transmission; the malleus, incus and stapes. Two muscles, chorda tympani nerve and the tympanic plexus of nerves.

Malleus: This bone is shaped like a hammer hence the name. This is the largest of the three ossicles of the middle ear cavity. It has a head, neck and three processes arising from below the neck. The overall length of the malleus ranges between 7.5 - 9 mm. Its head lies in the attic region of the middle ear effectively dividing the attic into an anterior portion and a posterior one. The anterior portion lies anterior to the handle of the malleus, while the posterior portion lies behind the handle of the malleus. During surgical procedures for attic

cholesteatoma clipping of this head will improve the exposure in the attic region. The head of the malleus on its posteriomedial surface has an elongated saddle shaped cartilage covered facet for articulation with the incus. This articular surface is constricted near its middle dividing the articular facet into a larger superior and a smaller inferior portions. The inferior portion of the articular facet lies at right angles to that of the superior portion. This projecting lower portion is also known as the cog or spur of the malleus. Below the neck the bone broadens and gives rise to the following: the anterior process from which a slender anterior ligament arises to insert into the petrotympanic fissure; the lateral process which receives the anterior and posterior malleolar folds from the annulus tympanicum, and the handle which runs downwards, medially and slightly backwards between the mucous and fibrous layers of the tympanic membrane. On the deep medial surface of the handle there is a small projection into which the tendon of the tensor tympani muscle inserts. Additionally the malleus is supported by the superior ligament which runs from the head to the tegmen tympani.

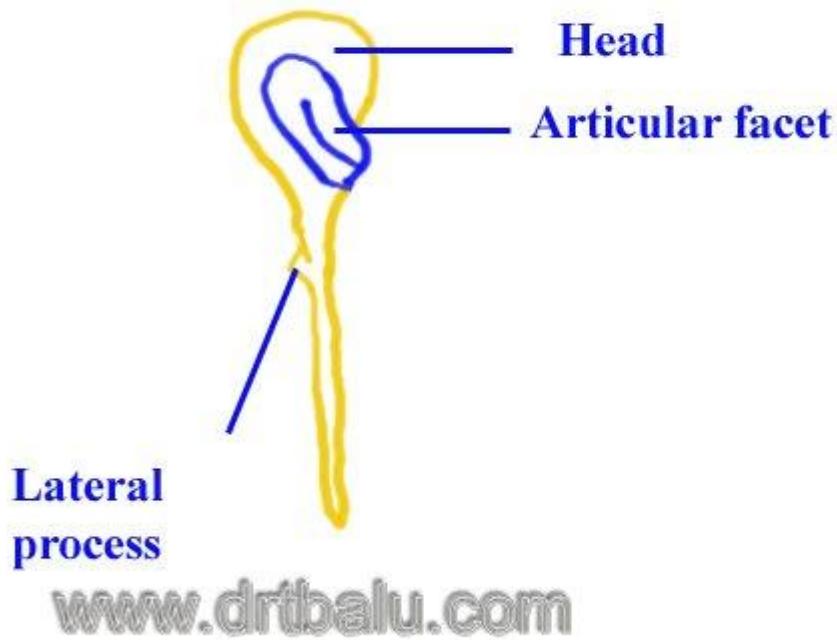
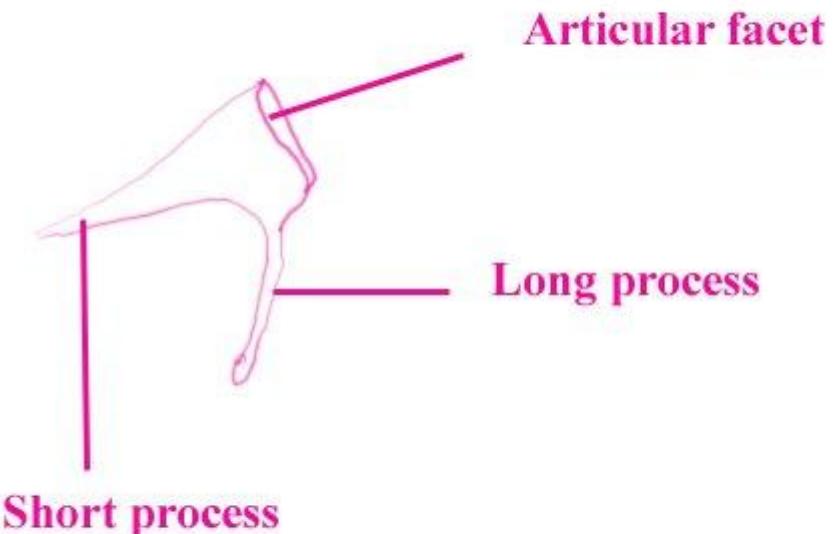


Figure showing malleus and its articular facets

Incus: This bone is shaped like an anvil. It articulates with the malleus and has a body and two processes. The body lies in the attic and has a cartilage covered articular facet corresponding to that of the malleus. The short process projects backwards from the body to lie in the fossa incudis. It is in fact attached to the fossa incudis by a short ligament. The long process of the incus descends into the mesotympanum behind and medial to the

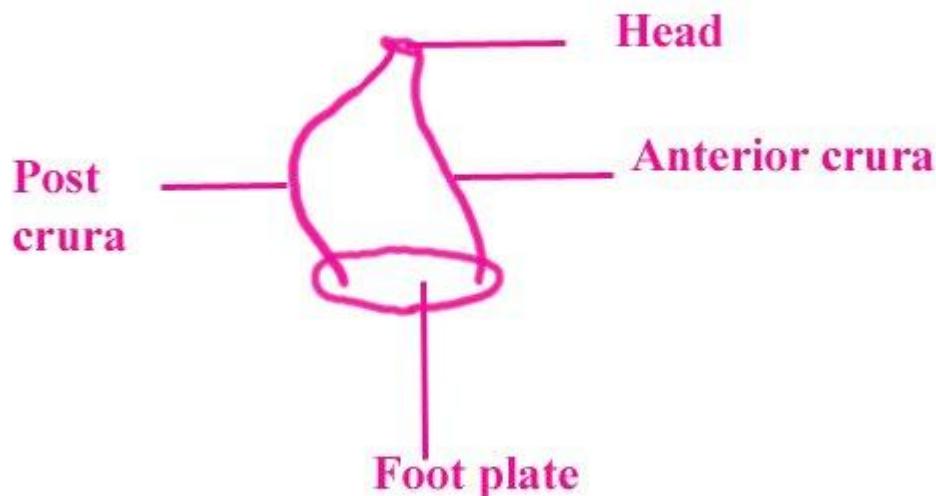
handle of the malleus. At its tip there is a small medially directed lenticular process which articulates with the stapes. The long process of the incus has precarious blood supply. This portion of the incus is prone for undergoing necrosis in disease conditions.



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Figure showing incus and its articular facets

The stapes: The stapes consists of a head, neck, two crura and a base (footplate). The head of the stapes points laterally and has a small cartilage covered depression for articulation with the lenticular process of the incus. The tendon of the stapedius muscle attaches to the posterior part of the neck and the upper part of the posterior crura. The neck of the stapes gives rise to two crura, the anterior crura is thinner and less curved than the posterior crura. The two crura join the foot plate which closes the oval window during life. The average dimensions of the foot plate are 3mm x 1.4mm. The long axis of the foot plate is almost horizontal, with the posterior end being slightly lower than the anterior.



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Figure showing stapes bone

Muscles of the middle ear:

Stapedius muscle: arises from the walls of the conical cavity within the pyramid. A slender tendon emerges from the apex of the pyramid and inserts into the stapes. This muscle is supplied by a small branch from the facial nerve. The stapedial tendon is inserted into the neck of the stapes. On contraction this muscle rocks the stapes backwards holding it firm against the annular ligament preventing excessive transmission of sound into the inner ear. This muscle has a protective role to play. It protects the inner ear from insults caused by loud noise. Patients with facial nerve palsy have hyperacusis because of lack of action of this muscle.

Tensor tympani muscle: This long slender muscle arises from the walls of the bony canal which lie above the canal for the Eustachian tube. Parts of the muscle also arise from the cartilaginous portion of the Eustachian tube and the greater wing of sphenoid. From these origins the muscle passes backwards into the tympanic cavity lying on the medial wall of the middle ear just below the level of the facial nerve. The bony covering of the canal is often deficient in its tympanic segment where the muscle is replaced by its tendon. This tendon enters the processus cochleariformis, turns at right angles inserting into the medial aspect of the upper end of the handle of the malleus. This muscle is supplied by the mandibular nerve by way of a branch from the medial pterygoid nerve, which passes through the otic ganglion without synapsing. This muscle tenses the tympanic membrane by holding the handle of the malleus thus helping the middle ear in better sound perception.

Chorda tympani nerve:

This is a branch of the facial nerve. It enters the middle ear cavity through the posterior canaliculus which is present at the junction of the lateral and posterior walls. It runs across the medial surface of the tympanic membrane between the mucosal and fibrous layers passes medial to the upper portion of the handle of the malleus. Here it lies above the tendon of the tensor tympani muscle, continues forwards and leaves by way of the anterior canaliculus placed within the petrotympanic fissure. It joins the lingual branch of the V nerve with which it is distributed to the anterior 1/3 of the tongue.

Tympanic plexus:

Is found over the promontory. It is formed by the tympanic branch of the glossopharyngeal nerve, carotico tympanic nerves which supplies the sympathetic component. The tympanic plexus provide the following branches:

1. Branches to the mucous membrane lining the tympanic cavity, Eustachian tube, mastoid antrum and its air cells
2. A branch joining the greater superficial petrosal nerve.
3. The lesser superficial petrosal nerve, which contain all the parasympathetic fibers of the IX nerve. This nerve leaves the middle ear through a small canal below the tensor tympani muscle where it receives parasympathetic fibers from the VII nerve by way of a branch from the geniculate ganglion. The full nerve passes through the temporal bone to emerge lateral to the greater superficial petrosal nerve on the floor of the middle cranial fossa, outside the dura. It then passes through the foramen ovale with the mandibular nerve and accessory meningeal artery to the otic ganglion. Post ganglionic fibers from the otic ganglion supply secretomotor fibers to the parotid gland by way of the auriculotemporal nerve.

The mucosal lining of the middle ear cavity is varies according to the location. The attic or the epitympanum is lined by pavement epithelium, while the middle ear proper is lined by cuboidal epithelium and the hypotympanum is lined by ciliated columnar epithelium.

Citation:

- 1.

Anatomy of middle ear by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
http://drtbalu.co.in/ana_mear.html

Presbyacusis

Synonyms: Deafness of the elderly, Age related deafness.

Definition: Presbyacusis is defined as a progressive bilateral symmetrical age related sensorineural hearing loss. The hearing loss is confined to higher frequencies.

Presbyacusis is an added problem for the elderly who have a tendency to compensate for their loss of vision through their intact sense of hearing. They even tend to get isolated and become a social recluse due to this problem.

Factors responsible for presbyacusis: Various factors have been postulated as causes of presbyacusis. They are:

1. Hereditary: Features like early aging of the cochlea and susceptibility of the cochlea for drug insults are genetically determined.
2. Atherosclerosis: May diminish vascularity of the cochlea thereby reducing its oxygen supply.
3. Dietary habits: Increased intake of fatty diet may accelerate atherosclerotic changes in old age.
4. Diabetes: May cause vasculitis and endothelial proliferation in the blood vessels of the cochlea thereby reducing its blood supply.
5. Noise trauma: Exposure to loud noise on a continuing basis stresses the already hypoxic cochlea hastening the presbyacusis process.
6. Smoking: Is postulated to accentuate atherosclerotic changes in blood vessels aggravating presbyacusis.
7. Hypertension: Causes potent vascular changes, like reduction in blood supply to the cochlea thereby aggravating presbyacusis.
8. Ototoxic drugs: Ingestion of ototoxic drugs like aspirin may hasten the process of presbyacusis.

Cochlear pathology seen in presbyacusis:

Depending on the pathology seen in the cochlea, 4 different types of presbyacusis have been identified. They are Sensory presbyacusis, neural presbyacusis, stria presbyacusis and cochlear conductive presbyacusis. The aging cochlea present disorders that are symmetric in paired ears; but the extent of involvement at the cellular level may be uneven. Hence presbyacusis can occur in 4 differing pathological types, or in combination thereof. A study of pure tone audiograms and cytococholeograms show:

1. Abrupt high frequency hearing loss (attributed to sensory cell pathology (loss)).
2. Flat threshold hearing loss (seen in cases with stria atrophy)
3. Diminished speech discrimination (due to loss of cochlear neurons)
4. Gradual descending audiometric pattern (due to inner ear conductive disorder)

Patients with presbyacusis uniformly have poor threshold for frequencies in 8 Khz range. In fact the threshold was as low as 60% in most of the patients.

Sensory Presbyacusis: is caused by loss of hair cells at the basal end of the cochlea. This commonly occurs in an aging cochlea. The area of involvement may extend to involve even the speech frequency area of the cochlea. These changes cause a rapid decrease in the threshold for high frequency sounds.

The earliest changes occurring in the cochlea is the loss of stereocilia, which can be identified only on electron microscopy. The second change to occur is distortion or flattening of the organ of corti followed by loss of supporting cells. Finally the organ of corti appears as an undifferentiated mound of tissue on a basement membrane. There is a gradual reduction in the number of outer hair cells in the elderly more so in the basal area of the cochlea. This occurs to a lesser extent at the apex of the cochlea. The apical loss of outer hair cells is seen only in individuals of more than 70 years of age. The loss of inner hair cells is less marked, but follows the same pattern as the outer hair cells.

The wear and tear pigment lipofuscin is known to accumulate in the apical cytoplasm of the hair cells. The lipofuscin is assumed to be a waste product of lysosomal activity.

Neural presbyacusis: Is caused by a loss in the population of cochlear neurons, but the end organs are still functional causing severe loss in speech discrimination. Pure tone thresholds are nearly normal. Gaeth used the term Phonemic regression to describe this phenomenon. Studies have shown that speech discrimination scores are slightly better in the left ear when compared to the right, this has been attributed to the left cerebral dominance becoming manifest due to the degenerative changes affecting the auditory pathway. The loss of cochlear neurons is the most consistent pathologic change seen in these patients. It has been calculated that loss of cochlear neurons occurs at the rate of 2,100 (Schuknecht) neurons every decade. There are roughly about 35,000 cochlear neurons in a normal ear. The loss of cochlear neurons may be genetically determined. The atrophy occurs throughout the cochlea, but is more pronounced in the basal turn of the cochlea.

Strial Presbyacusis : (Also known as Metabolic presbyacusis) Atrophy of stria vascularis is commonly seen in this condition. Hearing loss in these patients is insidious in onset occurring during the 3rd - 6th decades of life. It progresses slowly. The clinical feature that identifies this condition from the other types of presbyacusis is the presence of a flat or a slightly descending audiometric curve. These patients respond well to the amplification produced by hearing aids. This type of presbyacusis carries the best prognosis because of this feature. Takahashi demonstrated two types of atrophy in these patients:

Type I: a patchy type more severe in the apical and extreme basal regions of the cochlea.

Type II: A diffuse type often showing normal strial thickness with large intercellular spaces that may not be visible under light microscopy.

All 3 layers of stria are involved in various degrees. The loss of strial tissue may cause changes in the composition of the endolymphatic fluid causing further damage to the cochlear hair cells.

Pure tone audiometry shows a flat curve because the pathology involves the whole of the cochlea. Speech discrimination is preserved. This type of presbyacusis is considered by many to be familial.

Cochlear conductive (Mechanical) presbyacusis: This type of presbyacusis is differentiated from others by a linear descending audiogram. This is postulated to be caused due to stiffening of the basilar membrane of the cochlea. The thickening has been found to be more severe in the basilar turn of the cochlea where the basilar membrane is thin. Speech discrimination is average for the given frequency.

Mixed presbyacusis: Has been recently introduced to describe conditions which features of either two or all of the types of presbyacusis discussed above. These patients have been clubbed under this mixed category to account for their varied manifestations.

Presbyacusis is a diagnosis of exclusion. All the other causes of sensorineural hearing loss must be ruled out before declaring the patient to be suffering from presbyacusis.

Management: Amplification of the sounds with the use of hearing aids (with proper features) must be considered in all these patients. Administration of placebo drugs like neurotropic vitamins may make the patient feel something is being done to alleviate his problem. In rare patients cochlear implants may be considered.

Citation:

- 1.

Presbyacusis by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/presbyacusis.html>

Otitis externa

Synonyms: Infections of the external ear

Definition: Otitis externa is defined as infection / inflammation of the external auditory canal / auricle. It can range in its severity between mild infections to a more severe one. It is one of the most common diseases involving the external ear.

Classification:

Otitis externa is classified as follows:

1. Acute diffuse otitis externa (commonly caused by bacteria)
2. Acute localised otitis externa (commonly furuncle)
3. Chronic otitis externa
4. Eczematous otitis externa
5. Fungal otitis externa
6. Malignant otitis externa

Predisposing factors responsible for otitis externa:

Under normal conditions the skin lining the external auditory canal is well protected by its self-cleansing mechanism. In diseased conditions several factors may come into play in the pathogenesis of otitis externa.

1. Absence of cerumen: The cerumen plays an important role in the protection of the external canal. It protects the external canal from moisture. It also has anti-bacterial properties which helps in the protection of the external canal. The cerumen also lowers the pH of the external canal making it difficult for the bacterial pathogens to colonize.
2. Removal of cerumen by ear buds: is one of the common causes of otitis externa. The act of removal traumatises the skin lining of the external canal making it vulnerable to infections.
3. Frequent exposure to water: external canal when constantly bathed in water loses its ability to protect itself. The presence of water macerates the skin lining of the external canal and also increase the pH of the external canal making it more favourable for bacterial colonisation. This condition is common in swimmers.

Acute diffuse otitis externa:

This is also known as the swimmers ear. This is an inflammatory condition involving the external canal in a diffuse manner. This condition is common in swimmers because of the propensity for the external canal to be exposed to water for long durations. This exposure leads to maceration of the external canal skin, and also lowers the pH of the external canal providing an environment favourable to infections.

Main symptoms:

1. Itching in the external canal
2. Tenderness on palpation
3. Aural fullness rarely occur due to the reduction in size of the external canal lumen due to oedema
4. Rarely stenosis of the external canal may occur causing accumulation of debris and secretions

Common signs:

1. Erythema of the external canal
2. Oedema of external canal
3. Secretions from the external canal (weeping canal)
4. Pain on mastication
5. Pulling of helix in a postero superior direction cause pain
6. In advanced cases fever and lymphadenopathy may occur (pre and post auricular nodes may be involved)

Stages of acute diffuse otitis externa: (Senturia)

Preinflammatory stage: is characterised by intense itching, edema and sensation of fullness in the ear.

Inflammatory stage: may be divided into mild, moderate and severe.

Mild acute inflammatory stage: here the cardinal features are increased itching, pain, mild erythema and oedema of the external canal skin. At later stages exfoliation of skin with minimal amount of cloudy secretions may be seen in the external canal.

Moderate acute inflammatory stage: in this stage the itching and tenderness of the external canal intensifies. The external canal is narrowed due to oedema and accumulation of epithelial debris.

Severe acute inflammatory type: In this stage pain becomes intolerable to such an extent the patient may refuse to eat, the lumen of the external canal becomes totally obliterated due to oedema and accumulated epithelial debris. Otorrhoea may become purulent. In addition regional nodes may also be involved. Infections from the external canal may involve the parotid gland via the fissure's of santorini.

Common organisms involved: Psuedomonas aeruginosa and staphylococcus aureus are commonly cultured from the external canal of these patients. The normal commensals like staphylococcus epidermidis and corynebacteria are conspicuously absent.

Management:

The aim is twofold:

1. Resolving the infection
2. Promoting the external canal skin's recovery to its original state.

Firstly the canal is cleaned atraumatically by gentle suctioning and debridement under microscope. Topical hydrogen peroxide solution instilled will help the process of debridement.

A cotton wick dipped in I.G. paint can be inserted in to the external canal and allowed to stay for a day. This will reduce the external canal skin oedema and will increase the size of the meatus. Ear drops containing a mixture of neomycin and 1% hydrocortisone may be instilled as ear drops at least three times a day. In addition to the antibiotic and anti-inflammatory effects this drug reduces the pH of the external canal making it more resistant to the organisms.

In severe cases oral antibiotics and anti-inflammatory drugs can be resorted to. Quinolones are commonly used oral antibiotic.

Acute localised otitis externa: This condition is otherwise known as furunculosis or circumscribed otitis externa. This is a localised infection usually found to involve the lateral 1/3 of the external canal. It also has a propensity to involve the posterior superior aspect of the external canal. This is caused due to obstruction of the apopilosebaceous units found extensively in this area.

Trauma to skin in this area followed by infection is commonly attributed cause. The organism responsible is commonly staph aureus.

Symptoms:

1. Localised pain

2. Localised itching
3. Purulent discharge if the abscess ruptures
4. If oedema or abscess occludes the external canal hearing loss can occur.

Signs:

1. Erythema of the skin
2. Localised abscess formation

Management:

If the abscess is pointing it can be treated by incision and drainage. Oral antibiotics should be used. The preferred drug of choice is penicillin or first generation cephalosporins. Anti-inflammatory drugs can be used to reduce inflammation and pain.

These patients must be advised to cut their nails short and to keep their hands clean, since this is the commonest route of infection.

Chronic otitis externa:

This is a chronic infection / inflammation involving the skin lining of the external canal. There is thickening of the skin lining of the external canal due to persistent low grade infection / inflammation.

Symptoms:

1. Unrelenting pruritus
2. Mild pain
3. Presence of dry skin in the external canal

Signs:

1. Asteatosis (lack of cerumen)
2. Hypertrophic external canal skin
3. Presence of dry flaky skin in the external canal
4. Mild tenderness on ear manipulation
5. Rarely mucopurulent otorrhoea

Cultures from the external canal of these patients are highly unreliable because they would have been using various antibiotic drops to surmount the problem.

Management:

Involves extensive use of acetic acid ear drops. This helps to reduce the pH of the skin lining the external canal making it more resistant to bacterial infections. In intractable cases steroid drops can be tried. Antibiotic drops may not be useful in these patients.

Surgery is indicated in extreme cases. A canalplasty is performed to widen the external canal. The involved skin may be removed to be replaced by a split thickness graft.

Eczematous otitis externa:

This condition includes various dermatologic conditions involving the skin of the external canal. It may range from atopic dermatitis, contact dermatitis, seborrheic dermatitis, neuro dermatitis, infantile eczema etc.

This condition is characterised by intense itching, in fact this could be the only complaint of the patient. On examination, erythema of the external canal skin may be seen. There may also be associated scaling and oozing from the canal skin.

Success lies in the management of the underlying dermatologic condition.

Otomycosis:

It is also known as Fungal otitis externa. This is the commonest type of otitis externa in tropical countries. This condition is associated with increased ear canal moisture, or following treatment of otitis externa by prolonged use of topical antibiotics. The protective cerumen layer is absent in these patients. This condition is more common in diabetics.

Symptoms:

1. Intense itching
2. Pain when otitis externa is coexistent
3. Blocking sensation due to the presence of fungal balls

Signs:

1. Inflamed external canal skin
2. External canal tenderness
3. Fungal debris (black in case of aspergillus and white in the case of candida). Invariably the infection is mixed type.

Management:

The condition is managed by careful aural toileting to remove the fungal balls. The best way to remove fungus from the ear canal is by aural syringing. Antifungal ear drops of clotrimazole can be administered. If secondary infections are present oral antibiotics and anti-inflammatory drugs may be resorted to.

Citation:

1.
Otitis externa by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
http://drtbalu.co.in/otitis_ext.html

Chronic suppurative otitis media

Definition:

Chronic suppurative otitis media is defined as a chronic infection of the mucosa lining the middle ear cleft. Middle ear cleft include the eustachian tube, hypotympanum, mesotympanum, epitympanum, aditus and mastoid air cell system.

Types of chronic suppurative otitis media:

Chronic suppurative otitis media is of two types:

1. Tubotympanic disease (safe type)
2. Atticoantral disease (unsafe type)

Tubotympanic disease: This is also known as safe disease because it is bereft of any serious complications. The infection is limited to the mucosa and the antero inferior part of the middle ear cleft, hence the name. This disease does not have any risk of bone erosion. The discharge any will flow through a perforation present in the pars tensa portion of the ear drum. This perforation is usually surrounded by a rim of remnant ear drum or at least the annulus is intact. (Central perforation). The perforation is usually reniform (kidney shaped) because of poor blood supply to the affected portion of the tympanic membrane.

The infective activity of safe disease is related to the frequency of upper respiratory tract infections, the discharge tending to increase with increasing frequency of upper respiratory infections.

Aetiology:

1. Could be a sequelae to inadequately treated acute otitis media.
2. Acute suppurative otitis media causing persistent perforation which could be infected from bacteria in the external auditory canal. This condition is known as persistent perforation syndrome.

Microbiology of CSOM: In all varieties of CSOM the major organism found in the discharge are gram negative bacilli i.e. *Ps. aeruginosa*, *E. coli*, and *B. proteus*. These organisms are not commonly found in the upper respiratory tract, but they are found in the skin of external auditory canal.

Clinical features of tubotympanic disease:

1. The discharge in this condition is profuse and mucopurulent in nature.
2. The discharge is not foul smelling.
3. Since the infected area is open at both ends i.e. the Eustachian tube end and the perforation in the ear drum, the discharge does not accumulate in the middle ear.
4. The ossicular chain is not at risk in this type of disorder, the conductive deafness caused is due to the presence of perforation in the tympanic membrane and thickening of the tympanic membrane.
5. Conductive deafness may also be accentuated by thickening of round window membrane due to the presence of secretions. Hearing loss is usually about 30 - 40 dB.
6. These patients have poorly pneumatised / sclerosed mastoid air cell system. This feature has been attributed to repeated attacks of middle ear infections during childhood causing inadequate pneumatisation of mastoid air cell system. In patients with pneumatised mastoid air cell system repeated middle ear infections can cause sclerosis with evidence of new bone formation. Mastoids in these patients may be sclerotic.
7. Pain in the ear when present is always associated with otitis externa. This commonly occurs when the patient attempts to clean the ear off the purulent secretions with a ear bud or cotton tipped applicator.

Pathology of tubotympanic disease:

Pathological changes depend on the stage of the disease. The stages are as follows:

Acute stage: This is where the ear is actively discharging. The mucosa of the middle ear cavity is hypertrophied, and congested.

Inactive stage: This condition is characterised by dry perforation of ear drum, commonly in its antero inferior part, close to the Eustachian tube orifice. The middle ear mucosa is normal.

Quiescent stage: Perforation of ear drum is present, the middle ear is dry and mucosa may be normal or hypertrophied.

Healed stage: Here the perforation of ear drum has healed by formation of thin scar. There may even be tympanosclerotic patches / chalky deposits on the ear drum. The ossicular chain is invariably intact.

Tuning fork tests show:

Rinne - Negative on the affected side

Weber - Lateralised to the good ear

Absolute bone conduction test - Not reduced

Pure Tone audiometry show conductive hearing loss. The hearing loss is invariably under 40 dB.

Management of tubotympanic disease:

Conservative management:

If the disease is active - with active ear discharge

Aural toileting - must be done using dry cotton swabs.

Suction method can be used to suck out secretions from the external canal and the middle ear cavity. The only disadvantage of this procedure is the risk of noise induced deafness.

Syringing the affected ear with warm saline mixed with acetic acid (1.5%) can be used to syringe the affected ear. This solution not only clears the ear of its purulent secretions, it also helps to remove crusts if present. The presence of weak acetic acid has bacteriostatic effect.

Role of antibiotics in the management of tubotympanic disease:

Antibiotics can be administered depending on the culture report. The best route of administration is topical because the presence of a large central perforation enables adequate concentration of antibiotics to reach the middle ear mucosa. Ototoxic drugs are to be avoided because the increased vascularity present in the middle ear mucosa will cause easy absorption of the drug into the inner ear fluids causing sensori neural hearing loss. Ciprofloxacin can be administered topically.

Oral amoxicillin in adequate doses or penicillins in adequate doses may be beneficial.

Role of antihistamines and nasal decongestants: Is questionable. Their role is to decongest the nasal and naso pharyngeal mucosa, pharyngeal end of Eustachian tube. Since there is associated perforation of tympanic membrane, secretions don't tend to accumulate inside the middle ear cavity. Topical nasal decongestants should not be used for more than a week, because of their propensity to cause rhinitis medicamentosa.

Precautions:

1. The ear must be kept dry. This can be achieved by keeping the ears plugged when taking head bath. Swimming must be avoided till the perforation heals.
2. Pre-existing sinus infections if any must be treated aggressively.
3. Presence of focal sepsis in the throat (tonsils commonly) must be ruled out.

Surgical management:

1. Surgical management aims at correcting the causative problems if any.

The presence of deviated nasal septum must be corrected as this could predispose to chronic sinus infections.

If focal sepsis is identified in the tonsils and adenoid then adenotonsillectomy needs to be performed.

After eradicating the possible focal sepsis only attempt must be made to definitively treat the perforation. If the ear drum has managed to stay dry for more than 6 months myringoplasty can be performed. Temporalis fascia is used as grafting material because of its availability in close proximity, its thickness is more or less similar to that of normal ear drum. One other added advantage is its low basal metabolic rate.

If middle ear mucosa is wet and oedematous then cortical mastoidectomy should be resorted to if conservative management fails. Mastoidectomy can always be combined with myringoplasty in the same sitting.

Atticoantral type of disease (Unsafe type of disease):

This is termed as unsafe because dangerous intra cranial and extra cranial complications can occur, proving fatal to the patient. This disease spreads by erosion of the bony wall of the attic. Cholesteatoma is commonly present in this condition. This disease is commonly seen in sclerosed mastoid cavities. Presence of granulation tissue is also common in this disorder.

This condition mainly affects the attic region of the middle ear. This region is pretty crowded, with the presence of the head of the malleus and incus. Any disease process involving crowded portions tend to cause more complications. Bone erosion occurs due to the presence of osteitic reaction in the bone tissue.

Definition of cholesteatoma: Cholesteatoma is defined as a cystic bag like structure lined by stratified squamous epithelium on a fibrous matrix. This sac contains desquamated squamous epithelium. This sac is present in the attic region. Cholesteatoma is also defined as 'skin in wrong place'. Cholesteatoma is known to contain all the layers of skin epithelium. The basal layer (germinating layer) is present on the outer surface of cholesteatoma sac in contact with the walls of the middle ear cleft.

Theories of bone invasion by cholesteatoma:

1. Pressure theory - states that increase in the pressure caused by enlarging cholesteatoma cause bone erosion. Ischemia has been attributed as the cause in this theory.

2. Enzymatic theory: Inside the cholesteatoma are present multinucleated osteoclasts and histiocytes. These cells release acid phosphatase, collagenase and other proteolytic enzymes. These enzymes are known to cause bone erosion.

3. Pyogenic osteitis: Pyogenic bacteria may release enzymes which could cause bone resorption.

Types of cholesteatoma:

1. Congenital cholesteatoma
2. Primary acquired cholesteatoma
3. Secondary acquired cholesteatoma

Congenital cholesteatoma: is known to arise from embryonic cell rests present in the middle ear cavity and temporal bone. These cell rests are known to commonly occur in cerebello pontine angle and petrous apex. In fact congenital cholesteatoma is seen as a whitish mass behind an intact tympanic membrane.

Derlacki and Clemis laid down the following as criteria to diagnose congenital cholesteatoma:

1. The patient should not have previous episodes of middle ear disease
2. Ear drum must be intact and normal
3. It is purely an incidental finding
4. If discharge and ear drum perforation is present then it should be construed that congenital cholesteatoma has managed to erode the tympanic membrane.

Clinical features: The disorder is an incidental finding. The common location of congenital cholesteatoma is the antero superior quadrant of tympanic membrane, postero superior quadrant being the next common site of involvement. Anteriorly situated congenital cholesteatomas are known to affect the Eustachian tube function causing conductive deafness due to middle ear effusion, whereas posterior congenital cholesteatoma is known to cause conductive deafness due to impairment of ossicular chain mobility.

Staging of congenital cholesteatoma:

Staging as suggested by Derlacki and Clemis: They were the first to stage congenital cholesteatoma. They classified congenital cholesteatoma into

1. Petrous pyramid cholesteatoma

2. Cholesteatoma involving the mastoid cavity
3. Cholesteatoma involving the middle ear cavity.

Potsic suggested the following staging mechanism:

Stage I: Single quadrant involvement with no ossicular / mastoid involvement.

Stage II: Multiple quadrant involvement with no ossicular / mastoid involvement

Stage III: Ossicular involvement without mastoid involvement

Stage IV: Mastoid extension

Nelson's staging:

Type I: Involvement of mesotympanum without involvement of incus / stapes

Type II: Involvement of mesotympanum / attic along with erosion of ossicles without extension into the mastoid cavity

Type III: Involvement of mesotympanum with mastoid extension

Staging this disease will help in deciding the modality of treatment and in predicting the long term prognosis.

Acquired Cholesteatoma: can be divided into two types, primary acquired and secondary acquired cholesteatomas.

Primary acquired cholesteatoma: In this condition there is no history of pre-existing or previous episodes of otitis media or perforation. Lesions just arise from the attic region of the middle ear.

Secondary acquired cholesteatoma: always follows active middle ear infection which manages to destroy the ear drum along with the annulus. This type of destruction is common in acute necrotising otitis media following exanthematous fevers like measles etc.

Theories to explain pathogenesis of cholesteatoma:

Various theories have been postulated to explain the pathogenesis of cholesteatoma. They are:

1. Cawthrone theory: This theory suggested by cawthrone in 1963 suggested that cholesteatoma always originated from congenital embryonic cell rests present in various areas of the temporal bone.
2. Theory of immigration: This theory was suggested by Tumarkin. He was of the view that cholesteatoma was derived by immigration of squamous epithelium from the deep portion

of the external auditory canal into the middle ear cleft through a marginal perforation or a total perforation of the ear drum as seen in acute necrotising otitis media.

3. Theory of invagination: This theory was suggested by Toss. He theorised that persistent negative pressure in the attic region causes invagination of pars flaccida causing a retraction pocket. This retraction pocket becomes later filled with desquamated epithelial debris which forms a nidus for the infection to occur later. Common organisms known to infect this keratin debris are Psuedomonas, E. coli, B. Proteus etc.

Toss also classified attic retraction pockets into 4 grades:

1. Grade I: The retracted pars flaccida is not in contact with the neck of the malleus.
2. Grade II: The retracted pars flaccida is in contact with the neck of the malleus to such an extent that it seems to clothe the neck of the malleus.
3. Grade III: Here in addition to the retracted pars flaccida being in contact with the neck of the malleus there is also a limited erosion of the outer attic wall or scutum.
4. Grade IV: In this grade in addition to all the above said changes there is severe erosion of the outer attic wall or scutum.

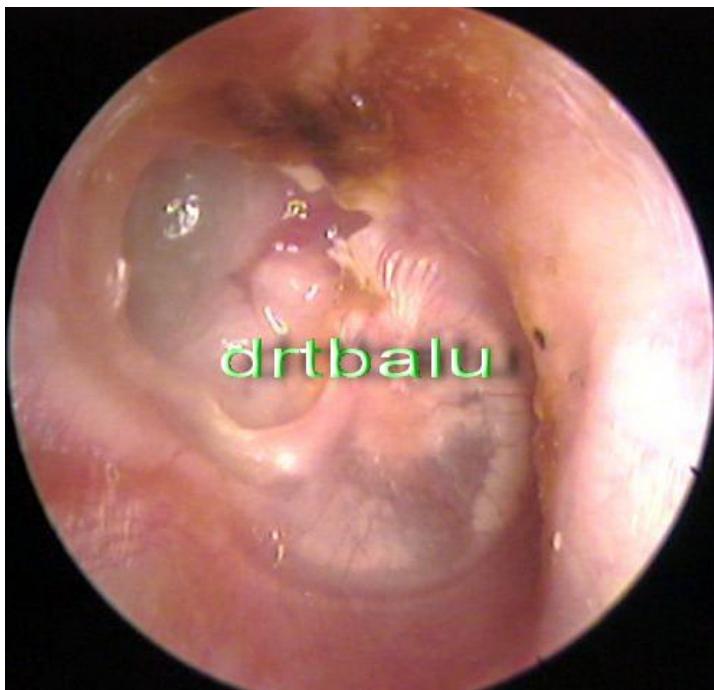
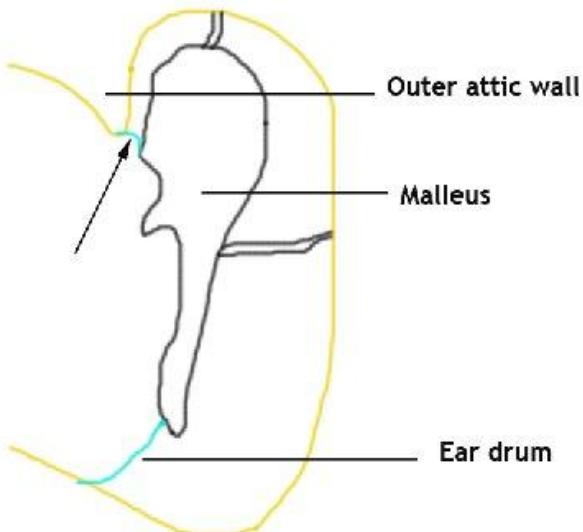


Figure showing attic perforation

Grade I retraction pocket:

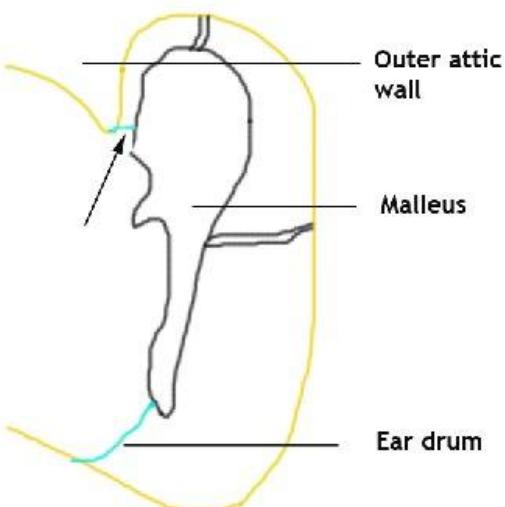
Grade I retraction pocket



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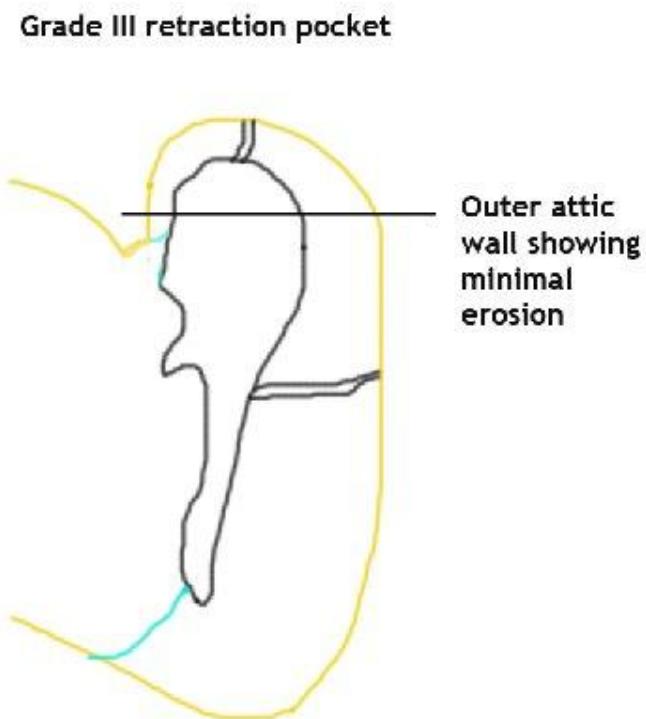
Grade II retraction pocket:

Grade II retraction pocket



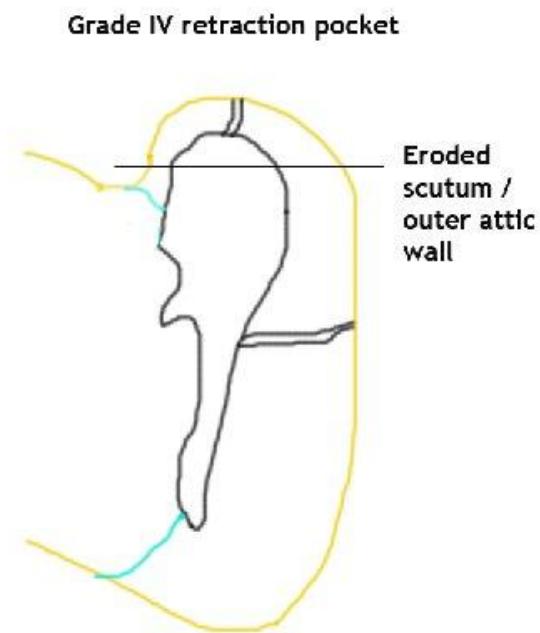
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Grade III retraction pocket:



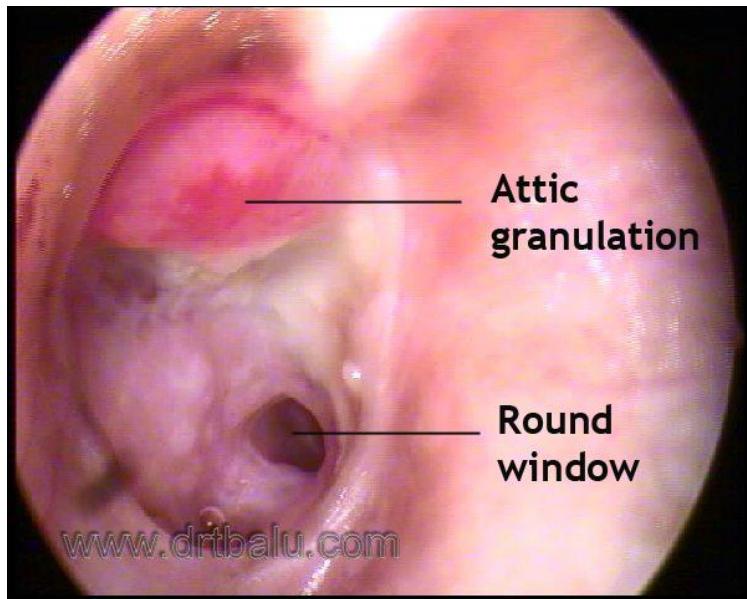
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Grade IV retraction pocket:



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4. Metaplastic theory: This theory was first suggested by Wendt in 1873. He took into consideration the histological changes seen in various portions of the middle ear cavity. The attic area of the middle ear cavity is lined by pavement type of epithelium. This epithelium undergoes metaplastic changes in response to subclinical infection. This metaplastic mucosa is squamous in nature thereby forming a nidus for cholesteatoma formation in the attic region.



Of all the above mentioned theories, the theory of invagination appears to be the most plausible one currently explaining the various pathologic features of cholesteatoma.

Clinical features of acquired cholesteatoma:

Ear discharge: is scanty and foul smelling. In fact the odour is best described as musty in nature. This is due to the presence of saprophytic infection and osteitis.

Hearing loss: is commonly conductive in nature. Some patients may even surprisingly have a normal hearing despite the presence of a huge cholesteatoma. This normal hearing could be attributed to the bridging effects of cholesteatomatous mass.

Sensorineural hearing loss if present could be attributed to the absorption of toxins through the round window membrane, or may be due to use of ototoxic antibiotics topically on a long term basis.

Ear ache: if present could be attributed to the presence of co-existing otitis externa, or presence of extradural abscess.

Tinnitus if present may indicate imminent sensorineural hearing loss.

Vertigo may be present if there is erosion of lateral semicircular canal by the cholesteatomatous matrix. Fistula test if performed is positive in these patients.

Fistula test: This test is positive if there is a third window is present in the labyrinth due to the erosion of the labyrinthine bone. This commonly occurs in the lateral semicircular canal area. This test is performed using a snugly fitting siegels pneumatic speculum and slowly

applying pressure by compressing the pneumatic bulb. If labyrinthine fistula is present the patient will feel giddy and will have nystagmus.

Facial palsy may indicate erosion of facial nerve canal with involvement of facial nerve.

On examination:

There is destruction of the outer attic wall, with presence of attic perforation. Cholesteatomatous flakes may be seen through the perforation like cotton wool.

There is associated sagging of the posterior superior meatal wall.

Hearing tests indicate conductive deafness commonly if labyrinth is uninvolved. It may turn out to be sensorineural hearing loss if there is associated erosion of the labyrinth.

X ray mastoids may show sclerosis with presence of cavity.

Management:

Since this is a surgical problem modified radical mastoidectomy is advocated in almost all of these patients.

The aims of the surgical procedure are as follows:

1. To exteriorise the disease
2. To create adequate ventilation to the middle ear cavity
3. To create a permanent skin lined cavity exposed to the exterior.

The various modifications of mastoidectomy procedures are discussed elsewhere.

Citation:

1.
Chronic suppurative otitis media by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/csom.html>

Stapedectomy

This surgical procedure is performed to treat deafness due to otosclerosis. Otosclerosis is caused by fixation of the foot plate of stapes which prevents efficient sound transmission to the oval window. The deafness caused is conductive in nature.

The surgical procedure is performed under local anesthesia. Advantages of performing this surgery under local anesthesia are:

1. Improvement in hearing can be ascertained on the table.
2. Bleeding is minimal under local anesthesia.

Indications for stapedectomy:

1. Conductive deafness due to fixation of stapes.
2. Air bone gap of at least 40 dB.
3. Presence of Carhart's notch in the audiogram of a patient with conductive deafness.
4. Good cochlear reserve as assessed by the presence of good speech discrimination.

Contraindications for stapedectomy:

1. Poor general condition of the patient.
2. Only hearing ear.
3. Poor cochlear reserve as shown by poor speech discrimination scores
4. Patient with tinnitus and vertigo
5. Presence of active otosclerotic foci (otospongiosis) as evidenced by a positive flemmingo sign.

Since a patient with otosclerosis is also an ideal candidate for hearing aid and surgery, the patient must be properly counselled regarding the advantages and disadvantages of both.

Anaesthesia:

Xylocaine with adrenaline mixed in concentration of 1:1000 is used to infiltrate the external auditory canal. 0.25 ml of the solution is infiltrated using a 27 gauge needle. Infiltration is given as illustrated in the diagram.

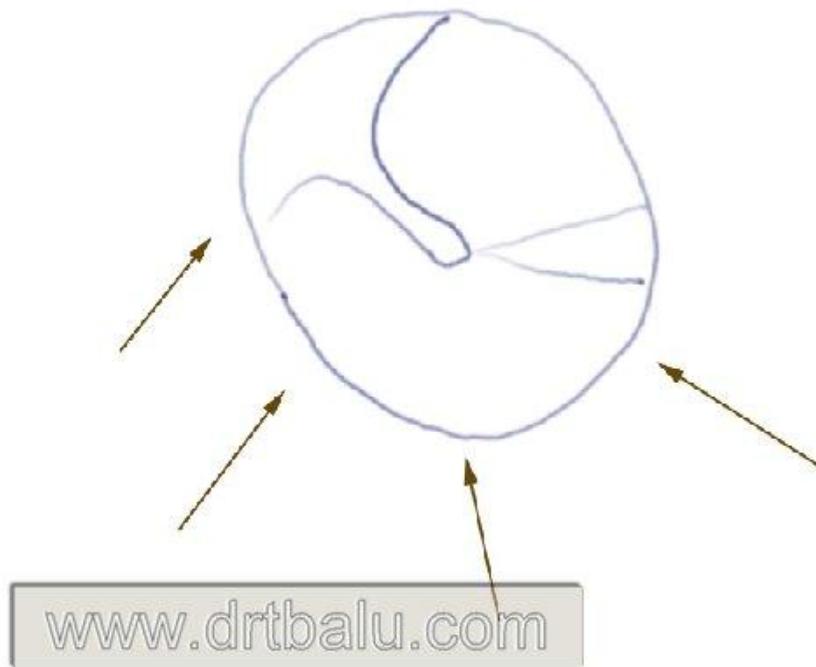


Fig showing the sites of infiltration of local anesthetic agent

Exposure: A large speculum is used to straighten the external auditory canal. A curved or triangular incision is made in the external canal skin beginning at 2mm away from the annulus. The incision extends from 11 o clock position to 6 o clock position as viewed in the right ear. The tympanomeatal flap is elevated up to the annulus. Using a sharp pick the annulus is slowly lifted from its groove, the middle ear mucosa is excised and the middle ear proper is entered.

Site of Incision



Fig showing incision made in the right ear

In most patients the posterior superior bony overhang must be curetted using a curette (designed by House). The long process comes into view. Curetting is continued till the base of the pyramidal process is visualised. Oval window is visualised. At this point round window reflex is tested by moving the handle of malleus and looking for movement of round window membrane. In otosclerosis this reflex is absent.

Using a hand burr a small fenestra about 0.6mm in diameter is made over the foot plate. The stability of the incus is left intact because the stapedial tendon is not cut at this point. From now on the steps may vary according to the surgeon's viewpoint. Some surgeons would like to insert the piston at this stage without disturbing the stability of the incus. The distance between the long process of incus and the foot plate is measured using a measuring rod. Appropriate size teflon piston is introduced and hung over the long process of the incus and is crimped after ascertaining whether its lower end is inside the fenestra. The stapedial tendon is cut at this point and the supra structure of the stapes is disarticulated and removed. The Tympanomeatal flap is repositioned.

Complications of stapedectomy:

1. Facial palsy

2. Vertigo in the immediate post op period
3. Vomiting
4. Perilymph gush
5. Floating foot plate
6. Tympanic membrane tear
7. Dead labyrinth
8. Perilymph fistula
9. Labyrinthitis

Citation:

1.
Stapedectomy by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://www.drtbalu.co.in/stapes.html>

Otosclerosis ¹

Synonyms: Otospongiosis, Ankylosis of foot plate of stapes.

Definition: Otosclerosis is a hereditary localised disease of the bone derived from the otic capsule characterised by alternating phases of bone resorption and new bone formation. The mature lamellar bone is removed by osteoclasia and replaced by woven bone of greater thickness, cellularity and vascularity.

History: In 1741 Antonio Valsalva described ankylosis of stapes while doing a post-mortem on the body of a deaf patient.

In 1894, Adam Politzer introduced the term "otosclerosis" and described the histopathology of the disease for the first time.

In 1912 Siebenmann introduced the term Otospongiosis to denote active otosclerotic foci.

Pathophysiology:

The primary pathological change occurs in the bony labyrinth with secondary effects upon middle ear and inner ear function. The otosclerotic focus may be asymptomatic, or if present in the area of foot plate of stapes it may give rise to ankylosis of foot plate with resultant conductive deafness. Otosclerotic foci may involve other portions of labyrinth causing sensori neural hearing loss and vestibular abnormalities.

Combinations of effects are possible in otosclerosis. They are:

Histological otosclerosis: Otosclerotic foci does not cause any symptoms and hence known as histological otosclerosis.

Stapedial otosclerosis: is the classical otosclerosis with fixation of stapedial foot plate causing conductive deafness.

Cochlear otosclerosis: The foci involve the cochlea causing sensorineural deafness.

Combined otosclerosis: Here in addition to fixation of foot plate of stapes there is also associated sensorineural hearing loss due to involvement of cochlea.

Otospongiosis: European otologists prefer to use this term to indicate the active phase of otosclerosis.

Incidence: Otosclerosis is common in Caucasian races. It is rarely found in Mongoloid and Negro population.

Sex incidence: In clinical practice otosclerosis is seen more often in women than in men. The ratio was found to be 2:1. Nowadays the authors believe the apparent female preponderance may be due to the fact that unilateral otosclerotic deafness is less common in women than in men. Noticeable deterioration in hearing also occur during pregnancy due to hormonal changes. Deafness due to otosclerosis may be initiated or made worse by pregnancy. **Causative factors / etiology:** Many theories have been proposed to explain the etiological factors of otosclerosis. They are:

1. Metabolic
2. Immune disorders
3. Vascular disease
4. Infection (Measles) currently accepted

5. Trauma : The petrous bone does not have regenerative capacity. This is because of the fact that the enzymes released during reparative phase are very toxic to the inner ear hair cells.

Pockets of tissue capable of regeneration may be sequestered in various portions of labyrinthine bone. These tissues could be activated by the presence of regenerative enzymes in the blood following bone fracture elsewhere in the body.

6. Temporal bone abnormalities (congenital)

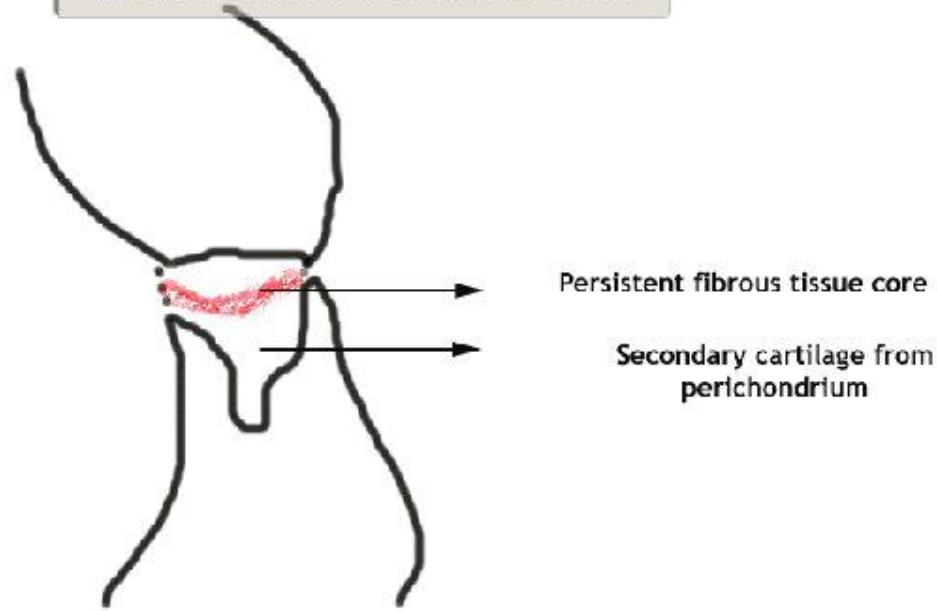
Genetic factors predisposing to otosclerosis: The tendency for otosclerosis to run in families has been documented. Authors have postulated an autosomal dominant mode of inheritance with varying degrees of penetration.

Otosclerosis is associated with osteogenesis imperfecta in 0.15 % of cases. This is known as Van der Hoeve syndrome or Adair - Dighton syndrome.

Sites affected by otosclerosis: The commonest site for appearance of otosclerotic bone is fissula ante fenestrum. This fissula is constantly seen in the labyrinthine capsule lying in front of the oval window. This area may contain unossified cartilage persisting even in adults. This area was referred to as Cozzolino's zone by Perozzi in memory of his teacher. Otosclerosis may occur in this area due to bony ossification of the cartilage. Residual cartilage may be present in the following areas of labyrinth:

1. Fissula ante fenestram
2. Fissula post fenestram
3. Intracochlear
4. Round window
5. Semicircular canals
6. Petrosquamous suture
7. Base of styloid process

In normal development the fissula appears as fibrous connective tissue bundle joining the vestibule with the tympanic cavity. This fibrous tissue is encased in primary cartilage which later gets replaced by bone. From the fissula the bone acquires a connective tissue lining which later becomes converted into perichondrium. The fissula is reduced in size by the production of new secondary cartilage from the perichondrium. These changes are completed by birth. The secondary cartilage remains throughout life as a stable, dormant cartilage and hence may even be considered as normal structure. It is only when this secondary cartilage gets ossified otosclerosis occur (Bast & Anson).



Diagrammatic representation of fissula ante fenestram

Otosclerotic changes may appear as a result of interaction between bone growth promoting substances circulating in the blood stream, and the unstable cartilaginous elements in the capsule of the labyrinth. Otosclerosis is often seen at times when the bone growth promoting substances are circulating in the blood as in pregnancy and following fractures of other bones.

Histopathology of otosclerosis:

The normal endochondral bone of labyrinthine capsule in which otosclerotic focus begins is compact in type. Ultrastructurally, lamellae composed of fine fibrils lying in a ground substance are concentrically disposed around haversian canals containing blood vessels and connective tissue. In otosclerosis there is sharply defined new bone formations that could be differentiated from normal bone by their deep carmine stain and by marked enlargement of bone spaces and haversian canals. The following are the changes which occur in a otosclerotic foci:

1. Focal / diffuse replacement of normal compact bone by irregular, loose cancellous bone with more deeply staining lamellae.
2. There is an associated increase in size of Haversian canals, cell spaces and marrow spaces with corresponding increase in vascularity. The blood vessels are frequently surrounded by a narrow margin of blue staining material that Manassee described first as Blue Mantle

zone.

3. Increase in osteocytes, and appearance of osteoblasts and osteoclast cells.

Histologically otosclerosis may be classified into:

1. Early focal otosclerosis
2. Diffuse active otosclerosis
3. Quiescent otosclerosis
4. Cochlear otosclerosis

Early focal otosclerosis: In this type the abnormalities are localised to one or two small areas of an otherwise normal foot plate section. The abnormal areas show an enlarged marrow space surrounded by a blue staining area on H&E staining.

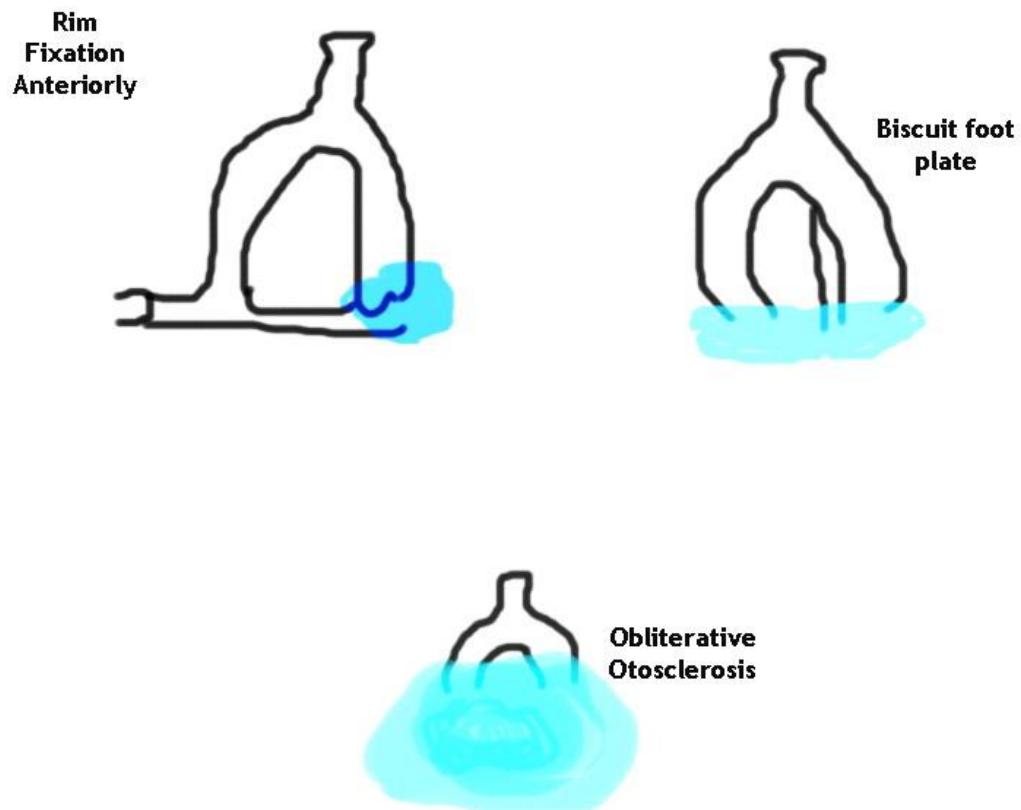
Diffuse active otosclerosis: In this type there is abnormal vascularity with a great increase in size and number of marrow spaces. Most of these spaces are lined by osteoblasts. In places around the circumference of the marrow spaces there is a scalloped appearance where bone has been recently absorbed. The numbers of osteocytes are greatly increased.

Quiescent otosclerosis: Here even though there may be some increase in the size and number of marrow spaces there is no evidence of bone formation or bone destruction. Osteoblasts and osteoclasts are only occasionally seen. This could be considered as a burnt out phase of the disease spectrum.

Cochlear otosclerosis: This condition causes pure sensorineural deafness without stapes fixation. Otosclerotic foci may occur in the otic capsule without the involvement of stapedial foot plate. The process of bone erosion and new bone formation which occur in otosclerosis releases enzymes like amylase, SGOT, SGPT etc which can enter into the endolymph via the round window membrane. These enzymes are toxic to the sensitive hair cells of the cochlea causing sensorineural hearing loss.

Clinical types of otosclerosis: Classification of various clinical types of otosclerosis is based on microscopic appearances of the diseased foot plate.

Rim fixation: Here the otosclerotic foci start from the anterior portion of the oval window niche. It gradually expands to involve the anterior portion of the foot plate causing fixation of the anterior portion of the foot plate only leaving the centre of the plate free.



Diagrammatic representation of various clinical types of otosclerosis

Biscuit foot plate: This type occurs less frequently. The focus originates in the foot plate itself and as it expands it gives rise to the biscuit or rice grain foot plate with delineated margins.

Obliterative otosclerosis: Rarely a large mass of otosclerotic new bone fills up the oval window niche obscuring the entire foot plate. This condition is known as obliterative otosclerosis. It is a difficult condition to manage surgically.

Clinical features:

Deafness: Typically deafness in otosclerosis is bilateral and gradually increasing in nature. Unilateral otosclerosis occurs in 15% of patients. Frequently it occurs between third and fifth decades of life. In majority of cases the deafness is conductive in nature. The deafness will not be noticed by the patient till the loss reaches 30 dB level. This is the deafness level in which understanding speech becomes difficult. These patients may hear better in noisy environment because the speaker has a tendency to raise his voice because of excessive ambient noise. This phenomenon a feature of otosclerosis is known as **Paracusis Willisii**.

In cochlear otosclerosis the deafness is purely sensorineural in nature. Some patients

may have both conductive and sensorineural hearing loss (mixed deafness) because of the tendency of bone reparative enzymes to damage the inner hair cells.

Patients with otosclerosis have characteristically quiet voice with good tone and the change in speech pattern may be detected only by close relatives.

Tinnitus: is a common symptom and occasionally could be the only presenting feature. The presence of tinnitus should alert the physician about the presence of cochlear otosclerosis. It could also be seen in some patients without cochlear degeneration due to abnormally increased vascularity of the otosclerotic bone. Mostly tinnitus indicates sensorineural degeneration. Tinnitus may be unilateral or bilateral. It is usually roaring in nature.

Vertigo: Transient attacks of vertigo are not uncommon in patients with otosclerosis. This could be due to the action of toxic enzymes released by the lesion into the vestibular labyrinth. These patients may even have coexisting Meniere's disease.

Clinical examination: The ear drum in these patients is normal (mint condition). Rarely during active phase of the disease the increased vascularity of the promontory may be seen through the ear drum. This sign is known as Flemingo's flush sign or Schwartz's sign. This indicates otospongiosis (active otosclerosis).

Hearing assessment can be done using tuning forks. For detailed description of tuning fork tests read the chapter titled clinical examination of the ear.

Pure tone audiometry will show precisely the amount and type of hearing loss. The presence of Carhart's notch is a classic audiometric feature in these patients. This Carhart's notch is present in bone conduction. There is a dip centred around 2000 Hz. This is actually an artefact. In cochlear otosclerosis audiometry reveals sensorineural hearing loss.

Stapes fixation causes an elevation in the bone conduction thresholds of 5dB at 500Hz, 10dB at 1000 Hz, 15 dB at 2000 Hz, and 5 dB at 4000 Hz. In the audiogram this creates a peculiar pattern known as Cookie bite audiogram. The bone conduction audiogram appears like a cookie having been bitten.

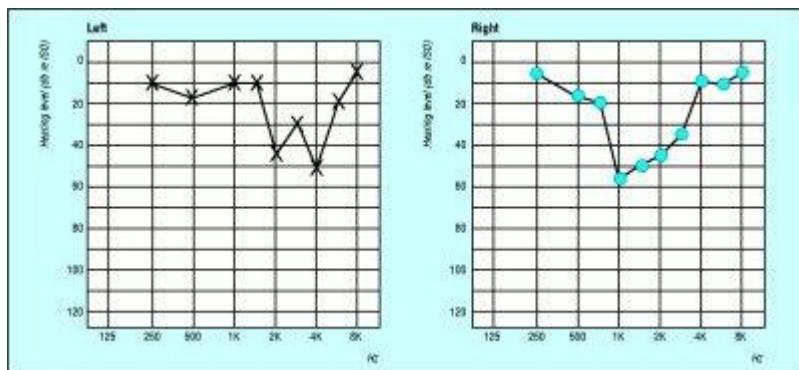


Fig showing a cookie bite audiogram

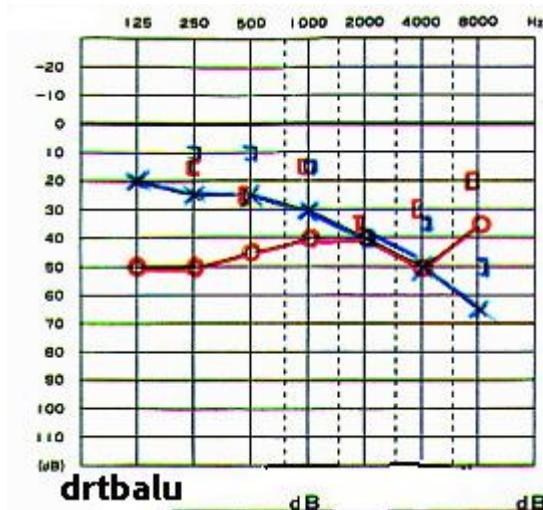


Fig showing audiogram with carhart's notch

Impedance audiometry is an useful investigation to diagnose otosclerosis. Middle ear compliance is often reduced. When stapes is fixed stapedial reflex is absent. The typical impedance curve is As curve.

All these patients with pure conductive deafness have excellent speech discrimination thresholds.

Management:

Medical: The aim of medical management is to convert an active otosclerotic foci into an inactive or quiescent foci. Fluride is the drug of choice.

Indications of fluride therapy:

1. Patients with surgically confirmed otosclerosis who show progressive sensorineural deafness disproportionate to age.
2. Patients with pure sensorineural loss with family history, age of onset, audiometric pattern and good auditory discrimination indicate the possibility of cochlear otosclerosis.

3. Patients with radiological demonstration by CT scan of spongiotic changes in the cochlear capsule
4. Patients with positive Schwartz sign.
5. Post op treatment: If patients are found to have an active focus during surgery, fluride therapy is prescribed for 2 years.

Contraindications of fluride therapy:

1. Patients with chronic nephritis and nitrogen retention
2. Patients with chronic rheumatoid arthritis
3. Patient who are pregnant / lactating
4. In children before skeletal growth has been completed
5. Patients who show allergy for the drug
6. Patients with skeletal flurosis

Flurides act on otosclerotic foci by reducing osteoclastic bone resorption with a corresponding increase in osteoblastic bone formation. Fluride also has antienzymatic action thereby it can neutralise the toxic enzymes released from the otospongiotic foci.

Dose: A daily dose of 50 mg of sodium fluride is given for a period of 2 years. In patients with positive Schwartz's sign the dose can be increased up to 75 mg per day.

Adverse effects of sodium fluride therapy:

1. Gastric disturbance
2. Arthritis
3. Skeletal flurosis

Surgical treatment: Stapedectomy

Hearing aids: These patients will benefit from the use of hearing aids if surgery is not acceptable to the patient or if it is risky. There is always a 1% risk of producing a dead ear during surgery even in the best of hands.

Citation:

1.
Otosclerosis - T. Balasubramanian - Google Books [Internet]. [cited 2011 Dec 31];Available from: http://books.google.co.in/books?id=IAE7-dVZKK8C&dq=otosclerosis&source=gbz_navlinks_s

Myringoplasty

Synonyms: Myringoplasty, Tympanoplasty.

Definition: Myringoplasty is a procedure used to seal a perforated tympanic membrane using a graft material.

Temporalis fascia is the commonly used graft material because:

1. It is an autograft with excellent chance of take
2. It is available close to the site of operation making its harvest easier
3. It has a low basal metabolic rate, brightening its success rate
4. Its thickness is more or less similar to that of tympanic membrane

There are two available methods of performing myringoplasty:

Overlay technique

Under lay technique

Overlay technique: This is a difficult technique to master. Here the graft material is inserted under the squamous (skin layer) of the ear drum. It is a difficult task peeling only the skin layer away from the tympanic membrane, placing the graft over the perforation and redraping the skin layer.

Underlay technique: This is a simpler and commonly used technique. Here the graft is placed under the tympanomeatal flap which has been elevated hence the name under lay. The major advantage of this procedure is that it is easy to perform with a good success rate.

Indications of Myringoplasty:

1. Central perforation which has been dry atleast for a period of 6 weeks.
2. As a follow up to mastoidectomy procedure to recreate the hearing mechanism

Prerequisites for myringoplasty:

1. Central perforation which has been dry for at least 6 weeks
2. Normal middle ear mucosa
3. Intact ossicular chain
4. Good cochlear reserve

Procedure: Firstly a temporalis fascia of adequate site must be harvested and allowed to dry.

The surgery is performed under local anesthesia. Temporalis fascia graft is harvested under local anesthesia conventionally and allowed to dry. The external auditory canal is then anesthetised using 2 % xylocaine mixed with 1 in 10,000 adrenaline injection. About 1/2 cc is infiltrated at 3 - o clock, 6 - o clock, 9 - o clock, and 12 - o clock positions about 3mm from the annulus. The patient is premedicated with intramuscular injections of 1 ampule fortwin and 1 ampule phenergan.

Step I: Freshening the margins of perforation - In this step the margins of the perforation is freshened using a sickle knife or an angled pick. This step is very important because it breaks the adhesions formed between the squamous margin of the ear drum (outer layer) with that of the middle ear mucosa. These adhesions if left undisturbed will hinder the take

up of the neo tympanic graft. This procedure will in fact widen the already present perforation. There is nothing to be alarmed about it.

Step II: This step is otherwise known as elevation of tympano meatal flap. Using a drum knife a curvilinear incision is made about 3 mm lateral to the annulus. This incision ideally extends between the 12 - o clock, 3 - o clock, and 6 - o clock positions in the left ear, and 12 - o clock, 9 - o clock and 6 - o clock positions in the right ear. The skin is slowly elevated away from the bone of the external canal. Pressure should be applied to the bone while elevation. This serves two purposes:

1. It prevents excessive bleeding
2. It prevents tearing of the flap

This step ends when the skin flap is raised up to the level of the annulus.

Step III: Elevation of the annulus and incising the middle ear mucosa. In this step the annulus is gradually lifted from its rim. As soon as the annulus is elevated a sickle knife is used to incise the middle ear mucosal attachment with the tympano meatal flap. This is a very important step because the inner layer of the remnant ear drum is continuous with the middle ear mucosa. As soon as the middle ear mucosa is raised, the flap is pushed anteriorly till the handle of the malleus becomes visible.

Step IV: Freeing the tympano meatal flap from the handle of malleus. In this step the tympano meatal flap is freed from the handle of malleus by sharp dissection of the middle ear mucosa. Sometimes the handle of the malleus may be turned inwards hitching against the promontory. In this scenario, an attempt is made to lateralise the handle of the malleus. If it is not possible to lateralise the handle of the malleus, the small deviated tip portion of the handle can be clipped. The handle of the malleus is freshened and stripped of its mucosal covering.

Step V: Placement of graft (underlay technique). Now a properly dried temporalis fascia graft of appropriate size is introduced through the ear canal. The graft is gently pushed under the tympano meatal flap which has been elevated. The graft is insinuated under the handle of malleus. The tympano meatal flap is repositioned in such a way that it covers the free edge of the graft which has been introduced. Bits of gelfoam are placed around the edges of the raised flap. One gel foam bit is placed over the sealed perforation. This gelfoam has a specific role to play. Due to the suction effect created it pulls the graft against the edges of the perforation thus preventing medialisation of the graft material.

Citation:

1.
Myringoplasty by drtbalu [Internet]. [cited 2011 Dec 31];Available from:
<http://drtbalu.co.in/myring.html>

Glomus jugulare

Synonyms: Paraganglioma, Chemodectoma, Ganglia tympanica, Vascular tumors of middle ear.

Definition: Glomus jugulare is defined as a collection of ganglionic tissue within the temporal bone in close relationship with the jugular bulb. The jugular bulb is closely related to the floor of the middle ear cavity (i.e. Hypotympanum).

History: Valentine in 1840 described this condition as ganglia tympanica. Guild recognised its histological relationship with the carotid body. Lattes and Waltner suggested that the ideal generic term for these structures is non-chromaffin paraganglioma.

Paraganglia cells are derived from the neural crest and are found widely distributed in the autonomic nervous system. Paraganglia having negative chromaffin reaction are termed non - chromaffin paraganglia. Guild in his anatomical studies on temporal bones found that on an average three glomus bodies were found in them. They were usually found in close relationship with the tympanic branch of glossopharyngeal nerve or the auricular branch of vagus. These bodies were supplied with non medullated sensory fibers from the adjacent nerves. They are supplied by branches from the ascending pharyngeal artery.

Even though the paraganglia cells are closely related to either the tympanic branch of glossopharyngeal nerve or the auricular branch of vagus, their position in the temporal bone is highly variable. Commonly they are found in the adventitial layer of the jugular bulb. In about 25% of cases they may be found over the mucosa of the promontory. Histologically, they resemble carotid body. It contains epitheloid cells interspaced in a highly vascular stroma of capillary and precapillary vessels. The proportions of the cellular and stromal components vary. Guild classified glomus tumors into two types depending on the amount of cellular and stromal components:

1. Cellular glomus bodies - when the cellular component is predominant
2. Vascular glomus bodies - when the vascular stromal component predominates.

Their sizes could be variable, but mostly they are ovoid in shape.

Paragangliomas of the temporal bone are generally divided into those that originate within the middle ear, glomus tympanicum tumors, and those that originate within the jugular fossa, glomus jugulare tumors. This latter term, however, is often used to refer to large tumors where the origin is difficult to determine. The predominance of the paraganglia within the jugular fossa likely accounts for the increased frequency of tumors with this origin. Classification systems that have been developed for temporal bone paragangliomas are used for staging purposes, surgical planning, and comparison among different therapeutic modalities.

Incidence: Glomus jugulare occurs in about 1 in 100000 patients. It is 6 times more

common in females when compared to males.

Hereditary pattern: It shows autosomal dominant inheritance with variable penetrance.

Endocrine activity: Even though these tumors are considered non chromaffin paragangliomas with no endocrine activity, some cases with endocrine activity by these tumors have been reported. It is hence important to look for evidence of endocrine activity by urine estimation of VMA (Vanillylmandelic acid).

Glomus tumors sometimes may show multicentric presentation i.e. present in both ears, or in conjunction with other paragangliomas. The carotid body being commonly the second site.

Pathophysiology: Glomus tumors are encapsulated, highly vascular, and locally invasive tumors. Inside the temporal bone they tend to expand along the pathway of least resistance such as air cells, vascular lumen, skull base foramina and Eustachian tube. They also invade and erode bone in a lobular fashion. The middle ear ossicles are commonly spared. Initially skull base erosion occur in the region of jugular fossa and postero inferior part of petrous bone. Later on extension occurs to the mastoid and adjacent occipital bone.

The parenchyma of the paraganglia consists of 2 primary cell types. Type I cells are more common and are typically round with indistinct cell borders. Type II cells are smaller and irregularly shaped.

Presentation: These tumors are slow growing, with very little symptoms. The diagnosis may easily be missed. Infact the average delay between the onset of symptoms and diagnosis varied from 6 years to 15 years. The first symptoms generally follow middle ear involvement is easily overlooked. Pulsatile tinnitus and conductive deafness are the common presenting symptoms. A red mass behind an intact ear drum (rising sun sign) may also be seen. In some 30% of cases cranial nerve palsies are common. Facial nerve is affected most commonly.



Otoscopic finding of a patient with glomus jugulare

Fig showing glomus jugulare (Rising sun sign)

Presenting features of Gomus jugulare:

1. Deafness - 69%
2. Middle ear mass - 75%
3. Pulsatile tinnitus - 55%
4. Imbalance - 8%
5. Otorrhoea - 5%
6. Facial palsy - 8%
7. Endocrine syndrome - 3%
8. Cranial nerve deficits
 - Hoarseness - 16%
 - Dysphagia - 16%
9. Headache - 15%
10. Visual disturbance - 6%
11. Presence of headache indicates intracranial extension
12. Dural sinuses may be involved may mimic sinus thrombosis

Clinical features: Otoscopic examination reveals a characteristic, pulsatile, reddish-blue tumor behind the tympanic membrane that often is the beginning of more extensive findings (ie, the tip of the iceberg). When the drum is examined under a microscope will show a pulsation of the reddish mass behind the drum. On seigalisation the mass blanches. This sign is known as *Brown's sign*. This is pathognomonic of glomus tumor.

Audiologic examination reveals mixed conductive and sensorineural hearing loss. The sensorineural component tends to be more significant with larger tumors.

Classification:

Glasscock - Jackson classification of temporal bone paraganglioma:

1. Type I : Small tumor involving the jugular bulb, middle ear and mastoid.
2. Type II: Tumor extending under the internal auditory canal. There may be intracranial extension.
3. Type III: Tumor extending into the petrous apex. There may be intracranial extension.
4. Type IV: Tumor extending beyond the petrous apex into the clivus and infratemporal fossa. There may be intracranial extension.

The Fisch classification of glomus tumors is based on extension of the tumor to surrounding anatomic structures and is closely related to mortality and morbidity.

Fisch classification:

1. Type A tumor - Tumor limited to middle ear (carries the best prognosis)
2. Type B tumor - Tumor limited to the tympanomastoid area with no infralabyrinthine compartment involvement
3. Type C tumor - Tumor involving the infralabyrinthine compartment of temporal bone with extension to petrous apex

This is divided into three types: C1, C2 and C3.

Type C1 - Tumor with limited involvement of the vertical portion of the carotid canal

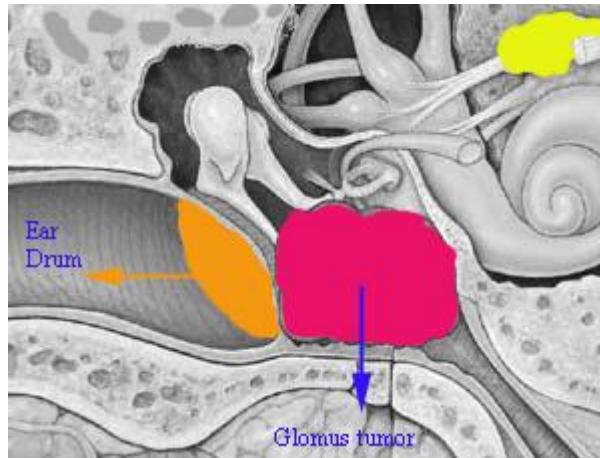
Type C2 - Tumor invading the vertical portion of the carotid canal

Type C3 - Tumor invasion of the horizontal portion of the carotid canal

4. Type D tumor has 2 types

Type D1 - Tumor with an intracranial extension less than 2 cm in diameter

Type D2 - Tumor with an intracranial extension greater than 2 cm in diameter

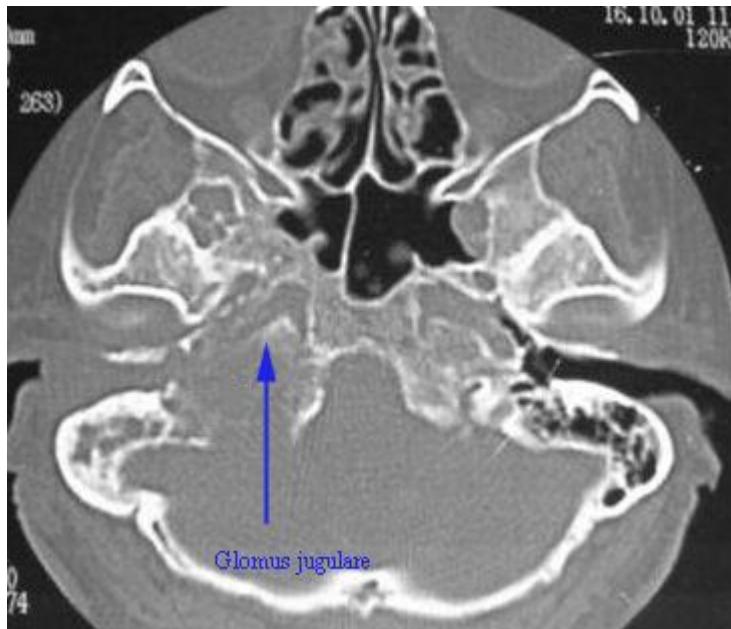


Diagrammatic representation of glomus tumor

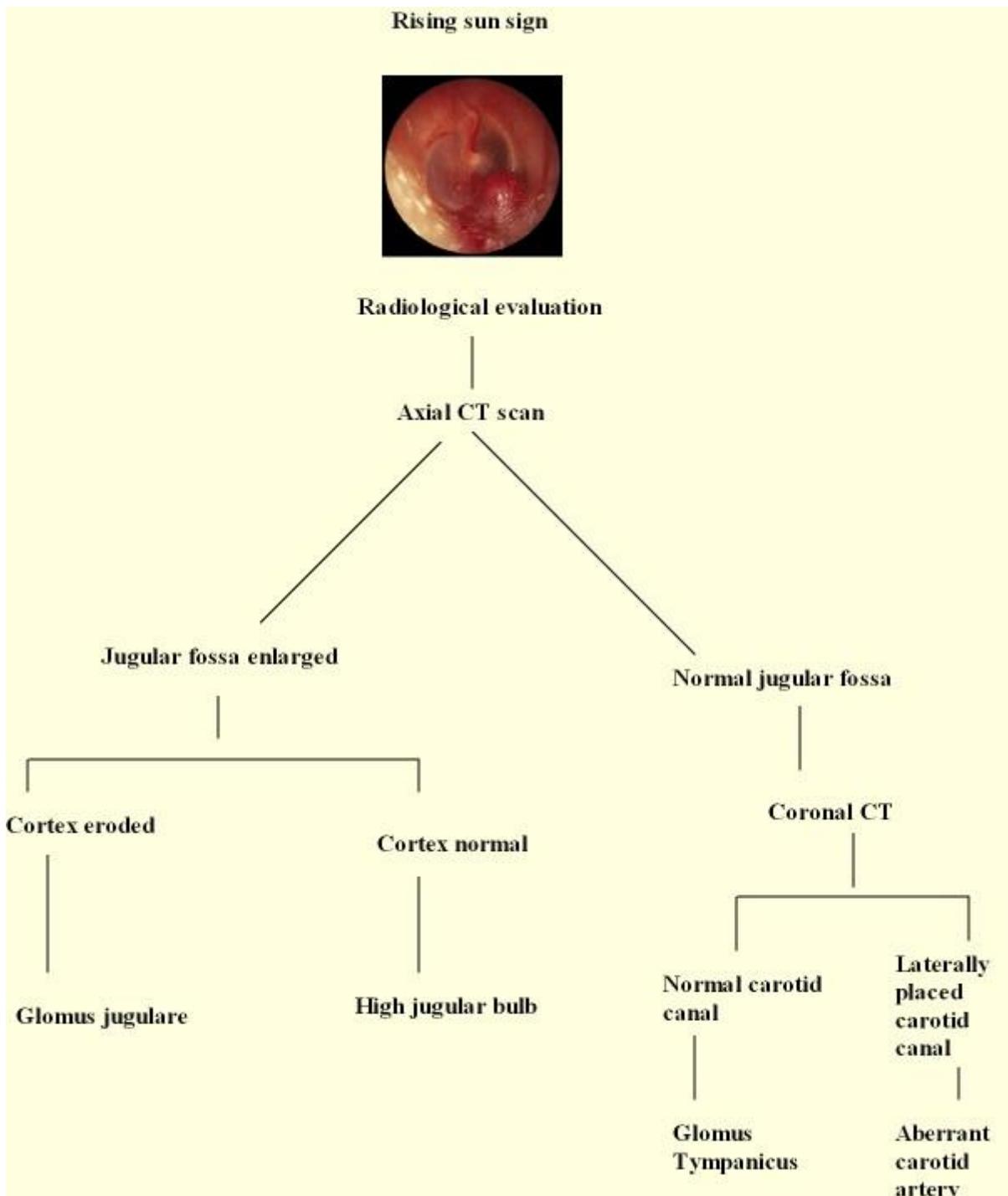
Investigations: Radiological investigations help in the diagnosis.

Plain X ray skull: May show enlargement of lateral jugular foramen and jugular fossa.

CT scan and Contrast MRI using Gadolinum enhancement is very helpful in delineating tumor extension.



CT scan showing glomus tumor



Flow chart showing radiological investigations to study glomus tumors in the middle ear

Applied anatomy of jugular bulb area:

The posterolateral portion of the foramen (pars venosa) contains the jugular bulb, posterior meningeal artery, and cranial nerves X and XI. The anteromedial portion (pars nervosa)

contains the inferior petrosal sinus and cranial nerve IX. The jugular bulb is situated between the sigmoid sinus and the internal jugular vein. The lower cranial nerves are situated medial to the medial wall of the jugular bulb. The inferior petrosal sinus enters the medial aspect of the jugular bulb via several channels anterior to cranial nerves IX, X, and XI. Many important structures are in proximity to the jugular bulb, including the internal auditory canal, the posterior semicircular canal, the middle ear, the medial external auditory canal, the facial nerve (posterior laterally), and the ICA (anteriorly) within the carotid canal. At the extracranial end of the jugular foramen, the ICA, internal jugular vein, and cranial nerves VII, X, XI, and XII are within a 2-cm area.

Treatment:

Treatment is mainly surgical. Complete resection of the mass is curative. Since it is a highly vascular tumor pre op intravascular embolization may help to reduce bleeding during surgery.

The particular surgical approach used to resect temporal bone paragangliomas depends on the location and extent of the tumor. Paragangliomas originating from the promontory of the middle ear and isolated to the mesotympanum can be resected by elevating the tympanic membrane and removing the tumor using microdissection techniques. If the tumor extends into the hypotympanum or the mastoid, a tympanomastoidectomy is performed and the tumor resected.

In extensive Fisch type 3 tumors the mass can be approached with help from neurosurgeons. The skull base approach ensures better exposure of the mass and facilitates complete resection.

Management of Fisch type 4 tumors is highly controversial. Irradiation of the mass has been tried with very little effect. Considering the slow growth rate of these tumors with a very long doubling time, these patients are best left alone with symptomatic treatment of the complications.

Complications of surgery:

Complications of surgery include death, cranial nerve palsies, bleeding, cerebrospinal fluid (CSF) leak, meningitis, uncontrollable hypotension/hypertension, and tumor regrowth.

Citation:

- 1.

Glomus jugulare by drtbalu [Internet]. [cited 2011 Dec 31]; Available from: http://drtbalu.co.in/glo_jug.html

Vertigo

Definition: Vertigo is defined as the subjective sense of imbalance. It has its root in Latin word "Vertere" which means to turn. Patient generally has difficulty describing his symptoms.

Synonyms: Vertigo, Dizziness, Unsteadiness.

Types of vertigo: Vertigo can be grossly classified into two types:

1. Rotatory: If the sensation is rotatory it is easy to describe it.
2. Non rotatory: There is obviously no sense of rotation and the patients have difficulty in describing this sensation.

These groups can further be subdivided into episodic and continuous according to the persistence of symptoms.

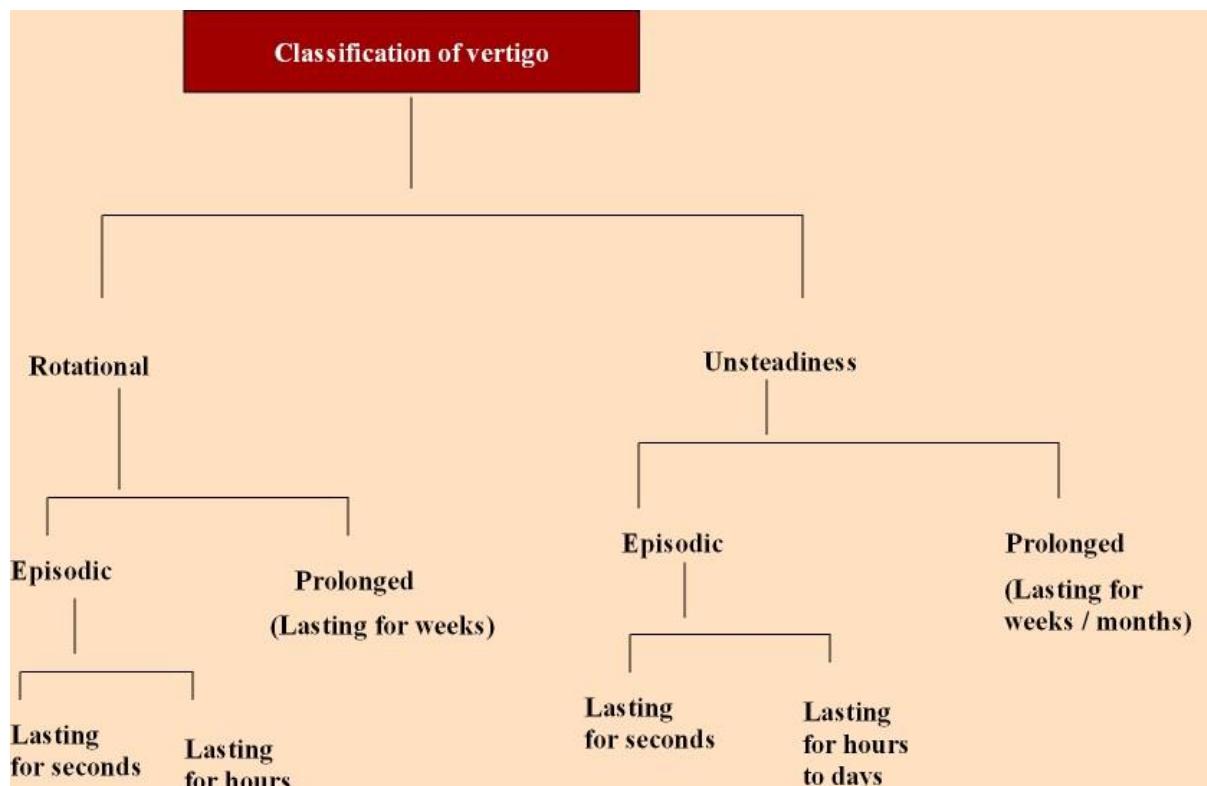


Figure showing classification of vertigo

Vertigo can be caused due to disturbances in the inner ear or in the central nervous system. It is important to differentiate the vertigo caused by peripheral and central causes.

Characteristic	Central	Peripheral
Severity	Mild	Severe
Onset	Gradual	Sudden
Duration	Weeks / Months	Seconds / Minutes
Positional	No	Yes
Fatigability	No	Yes
Associated symptoms	Neurologic / visual	Auditory

Table showing features of central and peripheral vertigo

A detailed history of the disorder is elicited from the patient with specific reference to postural variations, severity, mode of onset etc. The patient must also be quizzed about the associated neurologic / visual / auditory symptoms. Left to them the patient may not give the complete story, the examiner will have to take active control of history taking. The patients will be keen to emphasise the aspects which has impressed them the most. The patient should be specifically asked about periods of freedom from vertigo. History taking should be done chronologically.

A detailed history of drug intake is also a must to rule out drug induced dizziness / vertigo. A list of possible drugs are given below:

1. Anti Alzeimers - Drugs used in treatment of Alzeimers disease like *memantine, rivastigmine, tacrine* can cause dizziness.
2. Anti psychotics - Anti psychotic drugs like *chlorpromazine, prochlorperazine, fluphenazine, perphenazine, thioridazine, trifluoperazine* can cause dizziness.
3. Antidepressants - Can cause dizziness
4. Anxiolytics - Like *diazepam, alprazolam* can cause dizziness.
5. Mood stabilisers - Like *Gamapentin* can cause dizziness
6. Anticonvulsants - Like *Phenytoin* can cause dizziness

Examination of a Dizzy patient:

As a first step the peripheral causes of vertigo should be ruled out. This can be achieved by

a through otological examination. All the cranial nerves must be tested for integrity. Cerebellar function should be assessed by past pointing or by dysdiadokokinesis. Every dizzy patient must be examined for the presence of nystagmus with specific reference to its type. If the nystagmus is horizontal then in all probability the cause could be peripheral i.e. auditory. A direction changing horizontal or vertical nystagmus could indicate a central cause for the same.

Examination of corneal reflex: This test is sufficient to test the integrity of the trigeminal nerve. This test is so sensitive that it is positive even in patients with small acoustic neuromas.

Assessment if facial nerve: can be performed by just looking at the patient when he / she attempts to close the eyes. They will have inability to bury their eyelashes during tight closure of eyelids.

Performing a routine audiogram will assess 8th nerve function.

Cranial nerves 9th and 10th can be rapidly tested by looking for gag reflex. 11th nerve can be tested by asking the patient to shrug the shoulders, which is not possible if the nerve is affected. Hypoglossal nerve can be assessed by just asking the patient to protrude the tongue. It always tends to deviate towards the side of paralysis, in addition there may be wasting of tongue musculature on the side of the lesion.

Blood flow through the carotids should also be carefully examined to rule out carotid occlusion as a cause of giddiness. Patient's blood pressure and blood sugar levels must also be measured.

Nystagmus:

Presence of Nystagmus along with vertigo is always organic. Nystagmus is defined as a disturbance in ocular posture, characterised by a more or less rhythmical oscillation of the eye ball. The speed of the ocular movements may be the same in both directions or may be quicker in one direction when compared to the other. The direction of nystagmus is also important; a vertical nystagmus is more common in central causes of vertigo.

Basics of ocular movements: The movement of eyeball is controlled by 6 extraocular muscles i.e. 4 rectus and 2 oblique muscles. Among these muscles the superior oblique muscle is innervated by trochlear nerve and lateral rectus by abducens nerve. All the other muscles are innervated by oculomotor nerves. *There is something unique in the oculomotor supply, i.e. it innervates the ipsilateral ocular muscles except for superior rectus which receives its supply from the contralateral side.*

The superior oblique muscles are supplied by the contralateral trochlear nerve. The abducens nerve have two types of neurons:

1. Motor neurons innervating the ipsilateral lateral rectus muscle
2. Internuclear neurons supplying the contralateral medial rectus muscle.

Axes and planes of ocular movements: The axes and plane of ocular movements should be clearly defined before an assessment could be made. Three axes have been referred:

1. X axis : Also known as parasagittal or naso occipital
2. Y axis : Also known as transverse or interaural
3. Z axis : Also known as vertical axis

Movement along these axis are described as:

Torsional - Roll or rotation about X axis.

Vertical - Pitch or rotation about Y axis

Horizontal - Yaw or rotation about Z axis

Primary position of the eye: is the position of the eye in which there is pure horizontal or vertical rotation is associated with zero torsion. Clinically this position refers to the position of the eye when looking straight ahead with the body and head held straight and erect.

Secondary position of the eye: is reached by pure horizontal or vertical rotation of the eye.

Tertiary position of the eye: is reached when there is a combination of horizontal and vertical rotation from the primary position

Torsional eye movements: These are rotatory movements of the eye ball. When the eye ball rotates in such a way that the upper pole of the eye tilts towards the right of the subject it is known as clock wise rotation (always in the subject's point of view). When the upper pole of the eye tilts towards the left of the subject it is known as counter clock rotation.

The position of the eye ball is dependent on two sets of impulses, the visual and vestibular. The visual impulses are concerned with maintenance of position of eye in relation to the object of interest. The vestibular impulses help in maintenance of the position of eye ball in relation to that of the head.

Pendular nystagmus: is a horizontal to and fro movement of the eye ball. The to and fro movements are almost equal in velocity. This is commonly seen in patients with blindness. The eye ball is attempting to focus the image at the fovea of the retina. Since the patient is blind the eye ball keeps searching for visual object to focus hence this nystagmus. It is also known as the Blind Man's nystagmus.

Horizontal nystagmus: This is characterised by rhythmic oscillations in which the movement in one direction is significantly faster than the other. The slow movement is the pathological one while the faster one is the corrective component. The direction of the corrective faster movement is used to denote the direction of the nystagmus. The position in which the nystagmus is least marked is known as the *Null point*.

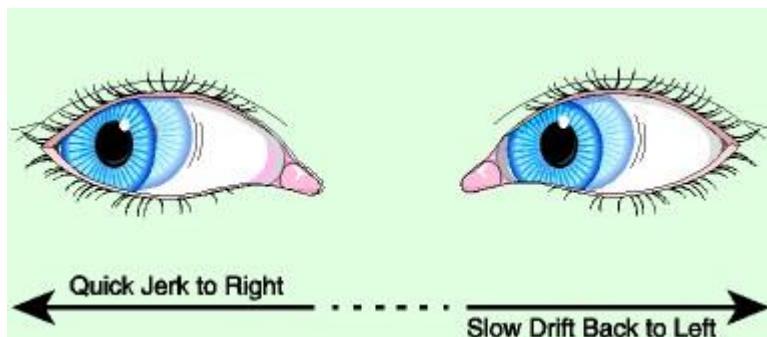


Figure showing horizontal nystagmus

Spontaneous nystagmus: is said to occur when the rhythmic movements are present on the forward gaze.

Induced nystagmus: is said to occur when the rhythmic movements of the eye are brought about by some specific test.

Gaze nystagmus: or intermediate nystagmus is said to occur during extremes of lateral or vertical gaze positions.

Procedure for examining for nystagmus:

1. The patient should be positioned in good light.
2. The object of focus should be at infinity
3. While looking for lateral gaze nystagmus care must be taken to ensure that the nose does not block the field of vision.

Causes for spontaneous nystagmus:

Labyrinthine: Labyrinthine nystagmus is usually biphasic. Its features are:

- a. It is usually associated with a sensation of vertigo
- b. It is always unidirectional.
- c. It is more marked when looking at the direction of the fast phase
- d. It is enhanced on removal of optic fixation or on eye closure. Frenzels glasses can be used to remove optic fixation. It is actually a 10 dioptre glasses worn by the patient. It removes optic fixation thereby enhancing labyrinthine nystagmus. These glasses make the eye look larger when viewed by the observer. Even small degrees of nystagmus will be noticed.

e. The nystagmus produced is fatigable

Central nervous system lesions causing spontaneous nystagmus: Nystagmus caused by lesions involving the central nervous system has the following features:

- a. Nystagmus is bidirectional (direction changing)
- b. Nystagmus could be commonly vertical
- c. Nystagmus usually is non fatiguing in nature
- d. Nystagmus is not enhanced on removal of optic fixation

Nystagmus caused due to drugs and toxins:

Drugs and toxins cause nystagmus by their effect on the central nervous system. The nystagmus induced by toxins and drugs more or less resembles that of central nervous system nystagmus

Ocular nystagmus: Any lesion affecting the macula, especially when the peripheral vision is still maintained may cause nystagmus which is pendular in nature. This nystagmus commonly occurs in amblyopia. Miner's nystagmus also falls into this category.

Congenital nystagmus: is often familial, usually horizontal and pendular. The null point in this type of nystagmus is close to the position of forward gaze. Closure of the eyes results in reduction of nystagmus.

Causes of induced nystagmus:

Nystagmus may be induced by clinical testing or by certain investigations. The most common clinical test inducing nystagmus is the positional test (Dix Hallpike manoeuvre). This is useful in diagnosing BPPV. This procedure is explained in detail under the topic BPPV.

Another way of inducing nystagmus is by performing a fistula test. This test is positive in the presence of labyrinthine fistula. In this test nystagmus and giddiness is induced by altering the pressure of air in the external canal.

Exposure to loud sounds in these patients also can cause nystagmus and giddiness. This is known as Tullio phenomenon.

In clinical setting nystagmus can be induced by performing caloric tests / cold caloric tests or by the use of optokinetic drum which can stimulate optokinetic nystagmus.

Oscillopsia: is the term used to describe the illusion of motion of environment due to an inadequate vestibulo ocular reflex. During head and body movements images are held

steady on the retina by the presence of vestibulo ocular reflex. Patients with reduced peripheral vestibular function often have oscillopsia. One method for testing for oscillopsia is to test the patient's visual acuity with a Snellen eye chart with the head held still, and repeating the same test during passive high frequency head movements produced by the examiner. The patient will have reduction in acuity if oscillopsia is present.

Investigations:

1. Audiometry
2. Caloric tests
3. Electric nystagmography
4. CT scan / MRI scan to rule out acoustic neuromas / CNS lesions

Causes of short lived episodic rotatory vertigo

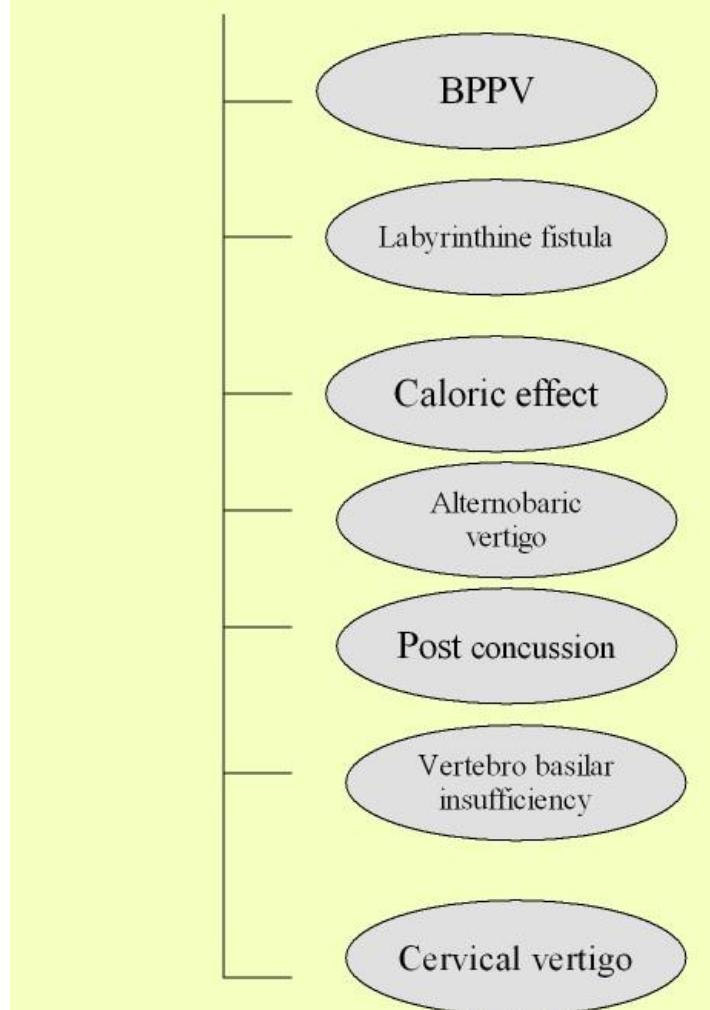


Figure showing causes of vertigo

Alternobaric vertigo is caused due to sudden change in altitude. This occurs in fighter pilots due to sudden altitude changes which occur during flying military jet fighter planes.

Episodic vertigo can also occur due to metabolic failure of labyrinth as seen in:

1. Meniere's disease
2. Syphilitic labyrinthitis
3. Delayed endolymphatic hydrops
4. Following middle ear surgery.

Destructive lesions of labyrinth: Can cause prolonged rotatory vertigo lasting for more than 24 hours, usually less than 3-4 weeks. These include:

1. Vestibular neuronitis
2. Trauma: Head injury, Ear surgery, Labyrinthectomy, and vestibular neurectomy
3. Labyrinthitis: Bacterial / viral
4. Vascular lesions
5. Metastatic deposits in CP angle

Unsteadiness: Patients who are unsteady may have problems describing their symptoms. Commonly it could be due to:

Physiological overload: Unsteadiness caused due to physiological overload of the vestibule or the central processing systems may cause unsteadiness. This could occur due to:

- a. Excessive input as can happen in a normal person with very rapid movements.
- b. May also occur as a result of abnormal input, especially from visual apparatus
- c. It may result from minor inadequacies in visual, proprioceptive or labyrinthine systems.

Unsteadiness lasting for hours / days may be due to temporary impairment of central vestibular connections or decompensation of vestibular system. Drugs are the most common cause for this temporary unsteadiness. Other causes include travels sickness, perilymph fistula, Active chronic suppurative otitis media, hyperventilation.

Vestibular inadequacy may cause unsteadiness lasting for weeks or months. This is often seen in elderly. Vestibulo toxic drugs like gentamycin can cause unsteadiness.

Vertigo following head injury may be due to:

- a. Post concusional syndrome
- b. BPPV
- c. Perilymph fistula
- d. Delayed hydrops

Treatment of vertigo:

Vestibular system can be suppressed by using labyrinthine sedatives like cinnarazine.

Wait for compensation to occur: In all peripheral causes of vertigo central compensation eventually occur in 6 weeks' time. Patient should be encouraged to performed labyrinthine exercises.

As a last resort the offending labyrinth can be eliminated by using intra tympanic drugs like streptomycin or surgical labyrinthectomy.

Citation:

1.
Vertigo by drtbalu [Internet]. [cited 2012 Jan 1];Available from:
<http://drtbalu.co.in/vertigo.html>

Examination of vestibular apparatus in infants and children

Introduction:

Examination of vestibular system in an infant or child is an arduous task. Adding to the obvious difficulties in clinical examination the child could be apprehensive and non-cooperative. In infants the process of examination could be very exacting. Vestibular dysfunction in children can present in a variety of ways ranging from a slight delay in acquiring head and postural control to acute episodes of vertigo and loss of balance. Since there is a close interaction between the vestibular, visual, proprioceptive, cerebellar and motor pathways in the maintenance of postural control and equilibrium in space it is very difficult to single out one faulty system as a reason for the defect.

History taking: A good history is a must before the actual examination of a dizzy child. It should concentrate on developmental milestones, history of drug intake.

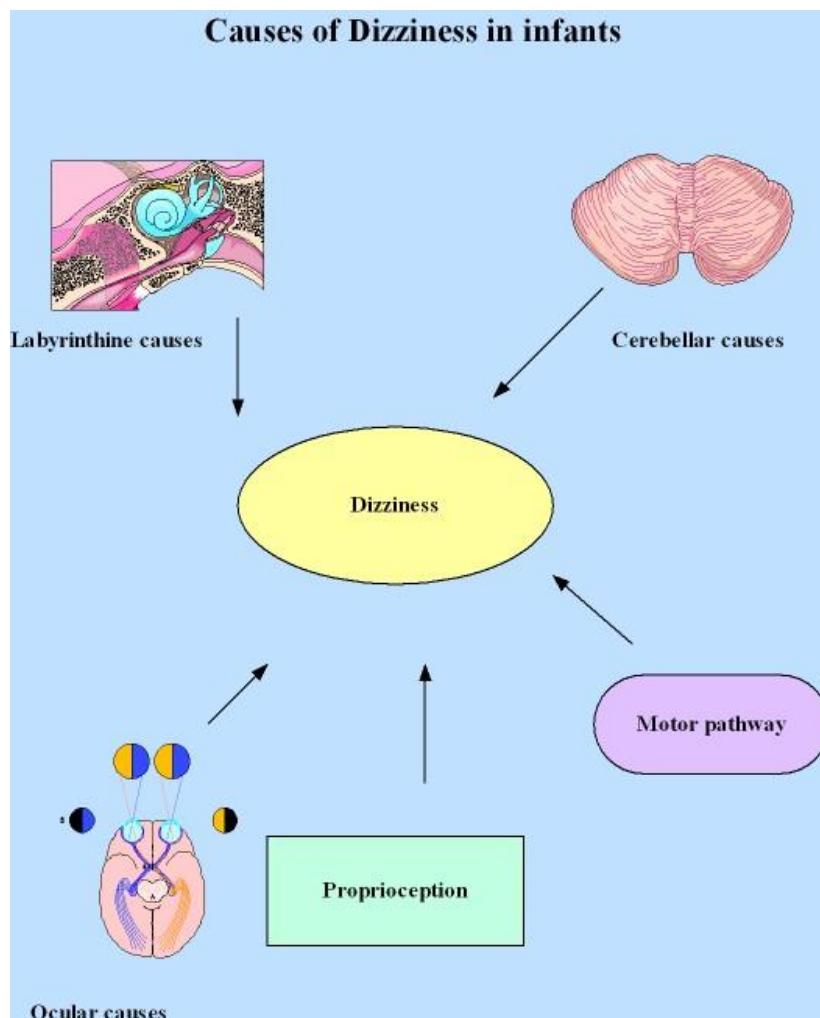


Figure enumerating various causes of dizziness in children

<ol style="list-style-type: none"> 1. Pregnancy: <ol style="list-style-type: none"> a. Intra uterine infections like toxoplasmosis, rubella b. Rh incompatibility c. Ingestion of ototoxic drugs d. Gestational diabetes e. Toxaemia of pregnancy 2. Neonatal period: <ol style="list-style-type: none"> a. Neonatal asphyxia b. Neonatal jaundice c. Respiratory distress syndrome d. CNS infections, sepsis e. Craniofacial anomalies 3. Developmental milestones: Sitting, standing, walking and speech 4. History of otitis, mastoiditis, head injury, CNS infections

Points to remember regarding development of inner ear:

1. Vestibular receptors are fully active and developed by 32 weeks of gestation. Hence Moro reflex can be theoretically elicited from this period till the child is 3 - 5 months of age.

2. Myelination of vestibular nerve fibers begins at about 16 weeks of gestation and is virtually complete at the time of birth.

To facilitate examination of a dizzy child, they are divided into 4 groups according to their age and levels of maturation of the central nervous system. They are:

Group I: Children under the age of 4 months are grouped here. In this group the tonic neck reflexes predominate. These reflexes can be demonstrated by passive or active motions of the head relative to the position of the body. This reflex is due to movement of endolymphatic fluid through the semicircular canals. These tonic neck reflexes are dependent on the integrity of vestibular and proprioceptive systems.

Neck righting: In this test active / passive rotation of head from the midline to one side

when the infant is lying supine will cause a rotation of the whole body in the direction of head turn.



Figure showing neck righting reflex test being performed

Asymmetric tonic neck reflex: This reflex is obtained with the baby lying supine with the head in midline position. Active or passive rotation of the head to one side while the infant's chest is restrained will produce flexion of the arms and legs on the side of the occiput with extension of arms and legs on the opposite side.

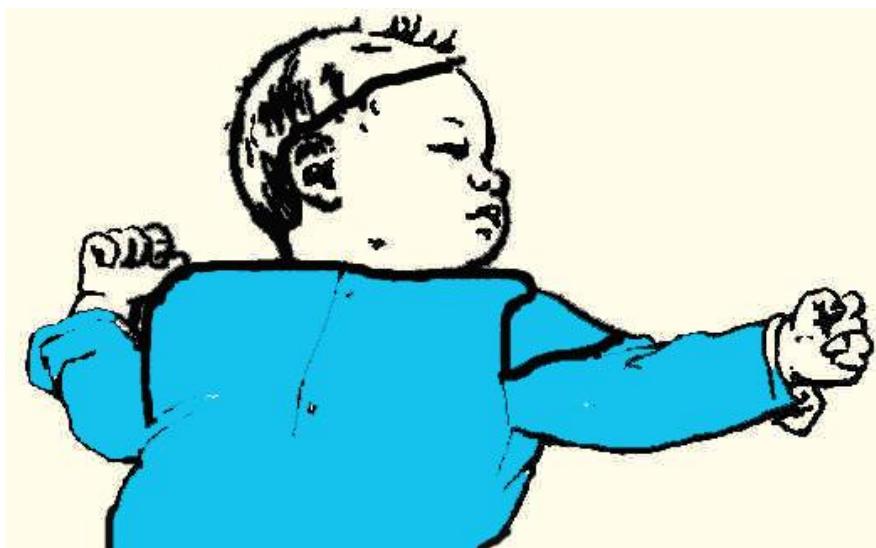


Figure showing asymmetric tonic neck reflex

Symmetric tonic neck reflex: This reflex has two stages. In the first stage, the baby is held in a horizontal prone position with the baby's chest in the examiner's arm or with the baby's chest on the examiner's lap. Dorsiflexion of the head will produce extension of the upper extremities and flexion of lower extremities.

In the second stage, abrupt ventroflexion of the head will produce flexion of the upper extremities and extension of lower extremities.

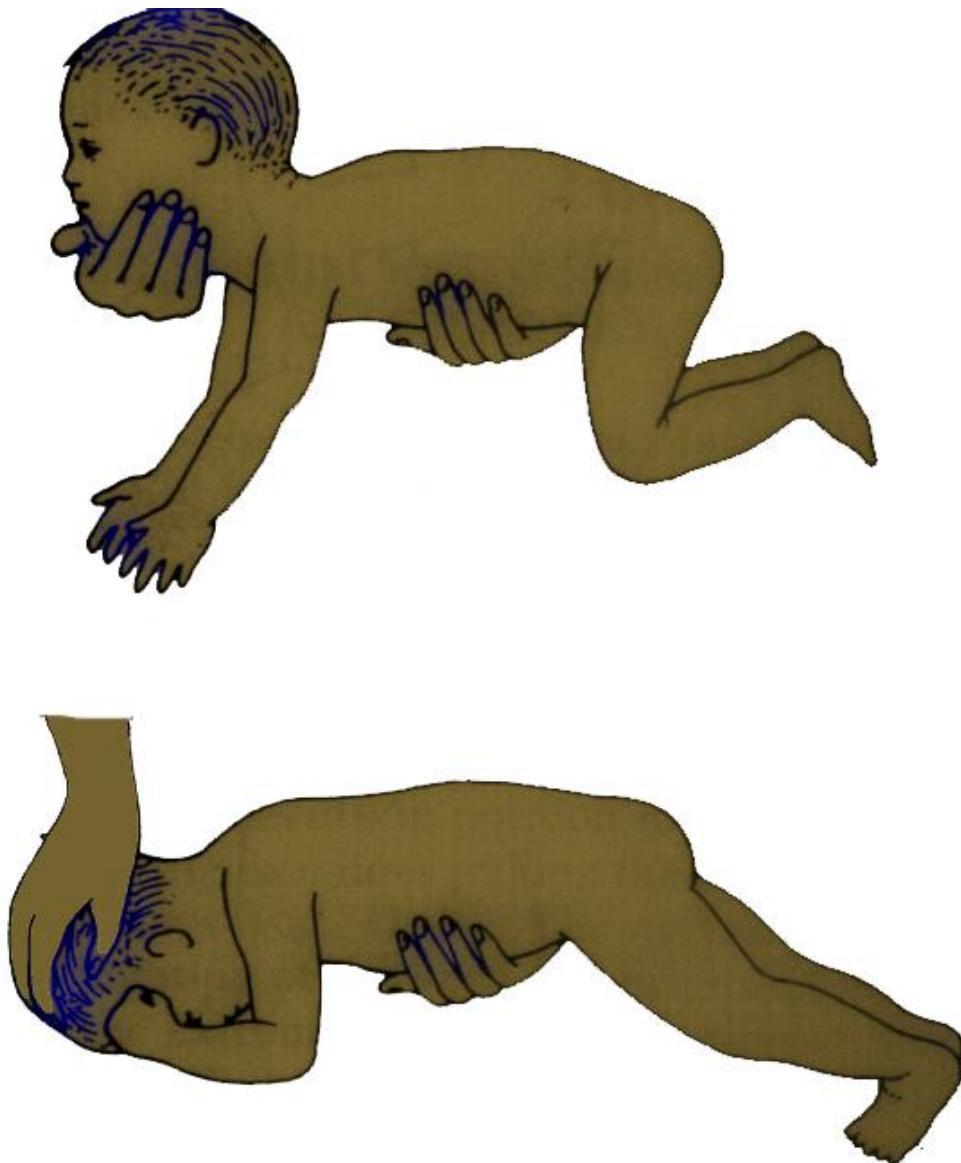


Figure showing symmetric tonic neck reflex

Moro reflex: For testing the Moro reflex the baby is made to lie in the supine position with the head ventroflexed and supported by the examiners hand. Now the head is abruptly dorsiflexed about 30 degrees in relation to the trunk. This movement will produce an extension and abduction of the arms, followed by an embrace. This reflex is otherwise known as startle reflex.



Figure showing Moro reflex being elicited

These tonic neck reflexes can be absent in the following situations:

1. Complete absence of labyrinthine control
2. In severely asphyxiated hypotonic child with severe central nervous system depression
3. Severe myopathic disorders

Vertical acceleration: In this test the baby is held in a supine position on the examiner's extended forearms. The head and trunk must be aligned and parallel to the ground. A rapid downward acceleration is produced to the baby's horizontal body by the examiner, who bends on his / her knees to a crouched position. A normal reflex response consists of abduction and extension of the arms with fanning of the hands. This response is similar to that of startle reflex (Moro's reflex). The only difference being the absence of dorsiflexion of the head, which eliminates proprioceptive input from the cervical vertebrae. Since the stimulus is vertical acceleration, it most probably stimulates the maculae of the utricle, as opposed to the cristae of the semicircular canals that are stimulated with the Moro reflex.

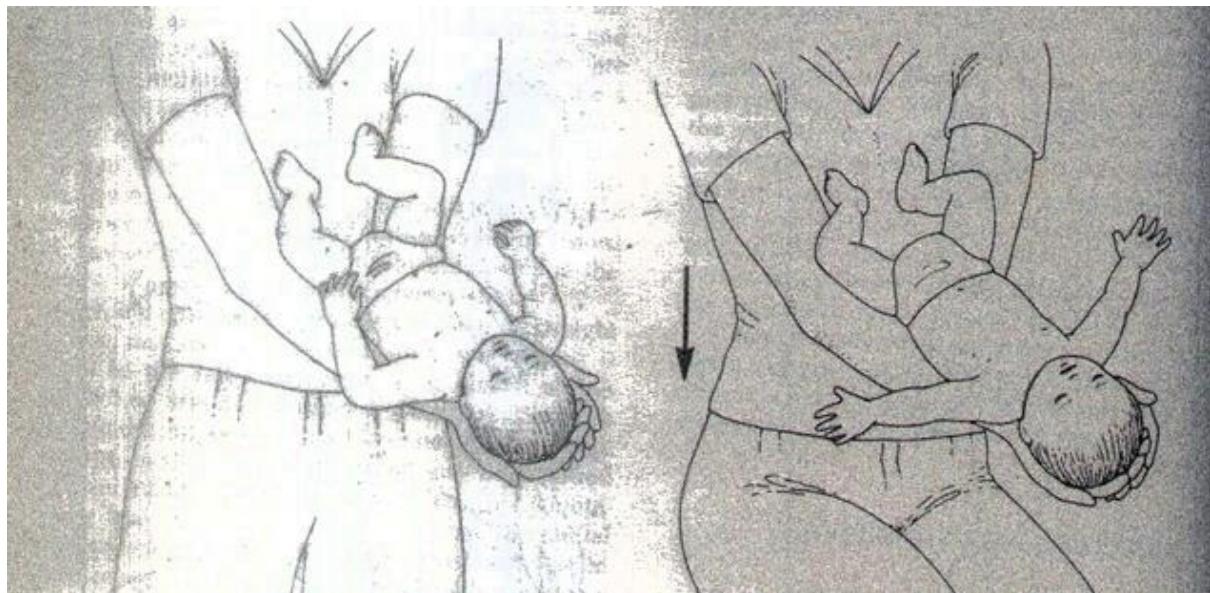


Figure showing vertical acceleration

Doll's Eye phenomenon: In testing for dolls eye phenomenon, the baby is held vertically under the armpits, with the head held 30 degrees forwards over the chest and is rotated for 360 degrees around an axis passing through the examiners head. Ten rotations in one direction are sufficient and provide a strong vestibular stimulus. The normal response is deviation of the eyes and head opposite to the direction of rotation. This phenomenon is present for first 2 weeks of life in full term neonates. Premature babies may have persistent dolls eye movement until 3 months of age. Gradually, the vestibular responses mature, nystagmus is superimposed, with a quick component in the direction of rotation.

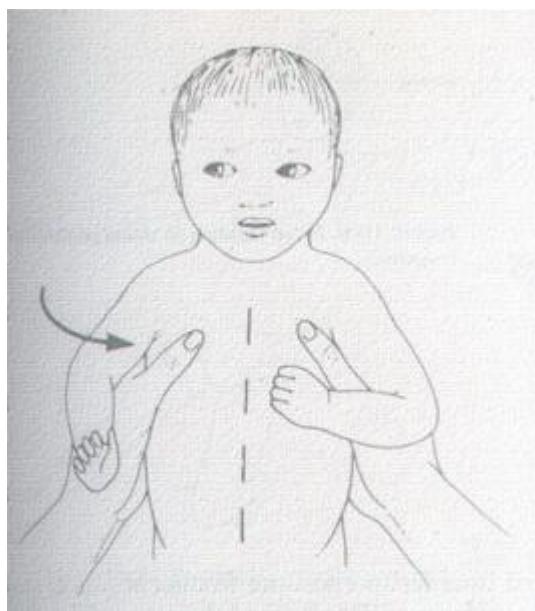


Figure showing Doll's eye movement

Group II: Babies of this group belong to the age group of 4 - 6 months. Babies in this age group vary in their developmental achievements. Many normal infants still have residual primitive tonic neck reflexes, while in others, righting responses will appear. Both these conditions are normal.

Group III: Babies of this group belong to 6 - 18 months. This is a period of rapid motor and sensory development. The pyramidal tract becomes myelinated. Integration of visual, labyrinthine and proprioceptive stimuli occurs during this phase. Righting reflexes are elicited by an abrupt tilt of the patient to change the patient's centre of gravity. Since the optical and vestibular righting responses are identical the baby must be tested blindfolded in order to eliminate visual cues. The most important of the righting reflexes is the head righting response. This can be obtained by picking up the infant from prone / supine position and bringing it to upright position by tilting the infant sideways, forwards or backwards. Every abrupt change of the head position in space will elicit vestibular head righting response. At the same time propping reactions of the extremities may be seen.



Figure showing righting reflex

Parachute reflex: This is also known as sentinel reaction. This is a basic protective body mechanism present throughout life. To test this reflex the baby is held under his / her arm pits with the back towards the examiner. The child is suddenly brought vertically down. The normal response consists of extension and abduction of arms, with extension of fingers as well as righting response of the head.

Hopping reaction: This reaction appears in normal full term infant by 8 - 10 months of age. The baby is tested in the standing position, with the examiner holding him or her around the chest and gently tilting him / her sideways, forwards or backwards. The general

response is initiation of a few steps in the direction of the tilt, accompanied by righting of the head. Acquiring of this reflex is a preparatory step to walking.

Equilibrium responses: These responses are more sophisticated and highly integrated righting reactions involving the whole of the body. This response can be tested in the sitting or kneeling position, with the examiner pulling the child by his or her arms sideways. The normal response consists of righting of the head and extension with abduction of the extremities on the side opposite to the direction of the tilt.

Electronystagmography: is a method of recording eye movements elicited by positional testing or during labyrinthine stimulation by rotation or caloric irrigation. This test is performed in a partially darkened room with the patient blind folded to eliminate fixation of gaze or optokinetic nystagmus. Microelectrodes are applied bitemporally for recording eye movements, and a neutral electrode is applied to the nasion. A position test is performed with the infant in supine, right lateral and left lateral positions. In older child a position test is also done in a sitting position.

Perotatory stimulus can be provided by commercially available torsion swing. Two perotatory stimulations are usually recorded with a five minute interval between two stimulations. The response obtained is a summation of responses elicited from both labyrinths.

In normal infants and children, the values for the speed of the slow component, amplitude and number of beats per torsion swing are identical for both right and left beating nystagmus. The total number of beats of nystagmus to the right and left are also identical. Directional preponderance is considered when the total number of beats in one direction exceeds by 25 percent the number of beats in the other direction. When directional preponderance is present it suggests vestibular dysfunction.

Ice - Cold caloric irrigation: Intra canalicular irrigation of cold water which is about 7 degrees less than that of body temperature stimulates the lateral canal. This test is ideally performed with the baby blind folded, in the supine position, with the head ventroflexed at 30 degrees. The child is also restrained. A ten second irrigation is a must for adequate stimulus. Recording should start immediately after the onset of irrigation. If the child is sleepy or irritable during the test the response may not be accurate. This test is a rather crude way of testing vestibular response to a stimulus. This test is hence performed only in cases of extreme doubts regarding the function of vestibular apparatus.

There is a maturation pattern in the development of caloric evoked nystagmus response. The amplitude and the number of beats increase in the first three months of life. The intensity of the nystagmus is directly proportional to the gestational age and the weight at birth. The latency of the response decreases with the gestational age and increasing birth weight.

Optokinetic stimulation: Optokinetic nystagmus can be evaluated in most children within three to six months of birth. As the child grows older, they learn to pay more attention to the moving images and better responses can be obtained in them. This nystagmus can be recorded in response to two speeds of rotation i.e. 3 degrees and 16 degrees per second. The frequency, amplitude and speed of the slow component can be analysed in response to the two rotational speeds. The information obtained is helpful in the evaluation

of overall quality of neurovestibular function.

Bithermal caloric testing: This test involves irrigation of the external canal for 30 seconds with water at 30 degrees and 44 degrees centigrade. A 10 minute interval is allowed between irrigations. This test is performed only in children aged 4 and older. The intensity of nystagmus represented by the speed of the slow component at the end point of nystagmus is used for calculations. It takes about 45 minutes for the procedure to be complete. Children younger than 4 years may not co-operate this long.

Jongkee's formula is used to calculate directional preponderance:

$$\frac{(R) 30 \text{ degrees} + (R) 44 \text{ degrees} - (L) 30 \text{ degrees} + (L) 44 \text{ degrees}}{X 100}$$

$$(R) 30 \text{ degrees} + (R) 44 \text{ degrees} + (L) 30 \text{ degrees} + (L) 44 \text{ degrees}$$

Citation:

1.
Examination of vestibular apparatus in children by drtbalu [Internet]. [cited 2012 Jan 1]; Available from: http://drtbalu.co.in/vesti_examc.html

Puretone audiometry

Synonyms: Audiogram, Assessment of hearing

Pure tone audiometry is used to measure the auditory threshold of an individual. The instrument used in the measurement of auditory threshold is known as the audiometer.

Audiometer: An audiometer has been described by the International Electrochemical Commission in 1976 as an instrument used for the measurement of acuity of hearing, and threshold of audibility. There are two types of audiometers widely used. They are:

1. Those that require a subjective response on the part of the patient and
2. Those that require no subjective response from the patient.

Examples include:

1. Pure tone audiometer is the classic example of the first type
2. Impedance audiometer / BERA are examples of the second variety

Components of a Puretone audiometer:

Oscillator: The role of the oscillator in a puretone audiometer is to generate electronically standardized frequencies within +/- 3% of their nominal value. The frequencies generated are 125, 250, 500, 750, 1000, 1500, 2000, 3000, 4000, 6000 and 8000 Hz.

Interrupter switch: The tones presented to the patient should be switched on and off. This feature is important because a continuous tone undergoes decay during a period of time. This switch gives the option of providing the tone in a continuous or an interrupted manner.

Equalisation circuit: This circuit contains resistors which helps in equalisation of the tones generated. This is because the threshold of human hearing is not uniform, the human ear is most sensitive at frequencies around 2 kHz. It is also insensitive at low or very high frequencies.

Output power amplifier: The signals produced by the oscillator needs to be amplified. The most important characteristic of the amplifier is that it produces very little distortion, and has a good signal to noise ratio. In most audiometers the power amplifier is run at constant high signal output levels.

Hearing level attenuator: The attenuator controls the level of the signal from the audiometer within the range of 110 - 120 dB. The attenuator can be varied in steps of 5

dB. The basic reference point is marked as 'O'. This indicates -5 to -10 dB hearing threshold levels. The attenuator steps should be accurate.

Output transducers: Is of three different types.

1. Ear phones
2. Bone vibrator
3. Loud speaker

The ear phones for audiometers are very special. They cannot be replaced or changed without calibrating the whole equipment. The pre requisites of a good ear phones are: 1. It should have a good long term stability
2. It should have a flat frequency response
3. It should be able to deliver high output sounds.

Bone vibrators: In contrast to ear phones bone vibrators have a limited dynamic frequency range. At low frequencies these vibrators show distortions.

Loud speakers: Are used in testing paediatric patients. It is used in free field audiology.

Calibration of the audiometer involves calibration of the audiometer proper, calibration of ear phones, and calibration of bone vibrators. The basic aim of these calibration procedures is to define the audiometric zero for the chosen earphone.

This can be performed using human volunteers or an artificial ear. The calibration of bone vibrators is the same as for earphones except for the measuring device which is different.

Pure tone air conduction testing:

This is a measurement of air conduction thresholds of audibility. Air conduction Threshold is in fact defined as the faintest tone a subject is able to hear via air conduction. In clinical setting pure tone audiogram is performed for two main purposes:

1. To assist in the diagnosis of ear pathology
2. To decide on the appropriate rehabilitation device which can be used to minimise the hearing disability.

Pure tone air conduction threshold is tested using head phones:



Figure showing a puretone audiometer



Figure showing headphones

Technique of measurement: Some audiologists assess the threshold of air conduction by going from an inaudible to audible stimulus intensity. This method is known as ascending method of estimation of threshold of hearing, while others assess the threshold of air condition by going from an audible to inaudible stimulus intensity. This is known as descending method of threshold estimation.

Instructions to the patient: The patient is instructed to raise the index finger if the sound is heard. The patient should respond even if the sound is faintly heard.

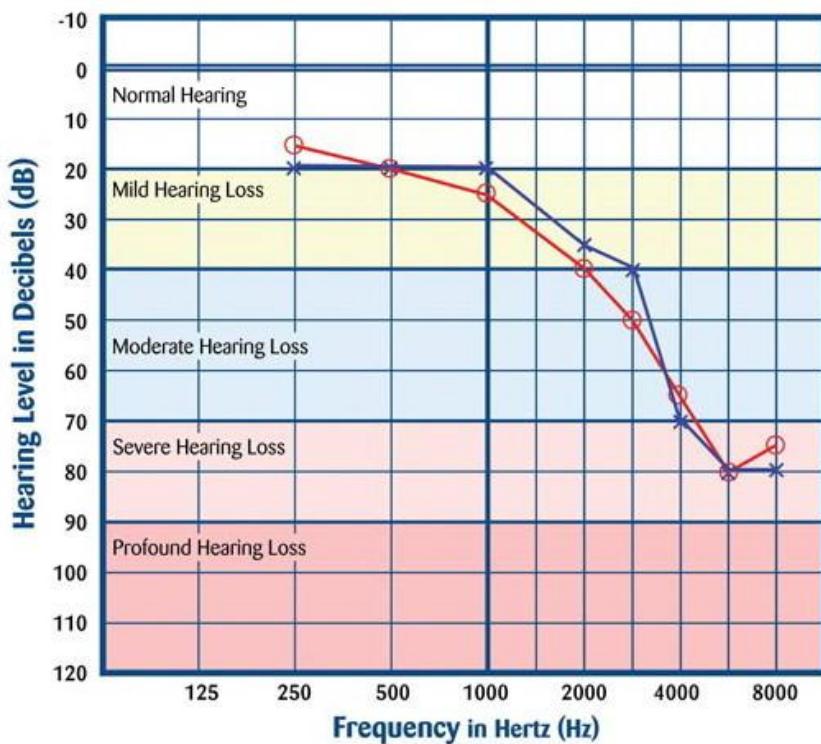
The head phones should be properly seated over the external auditory canal. This step should be performed with care because the patient's pinna comes in various shapes and sizes. Improper placement of head phones will cause threshold variations of even 15 - 20 dB.

The audiometer should be properly checked before performing the test. After the audiometer has had a warm up period, the tester should first place the ear phones on his own ears and listen to various frequencies and intensities of test tones. He should also listen to the masking noise, check for any audible clicks of the interrupter switch. Before placing the ear phones on the patient, the patient's ear should be examined for the presence of wax. If wax is present it should first be cleaned before the test could be performed. If the ear canal is small or tends to collapse when pinna is pressed, the test could not be valid. If the ear canal tends to collapse when pressure is applied to the pinna, plastic tubes can be inserted into the external canal to prevent such collapses.

To plot the recordings, red color ink is used to plot values of right ear, and blue color ink is used to plot values of left ear.

AUDIOGRAM

Left Ear  Right Ear 



Audiogram showing air conduction threshold

"Up 5-down 10" method of threshold estimation: This technique is based on Hughson - Westlake ascending technique.

Tones of short duration are used for threshold estimation. This method of threshold estimation involves the following steps:

Step I : The better ear is tested first in order to determine the need for masking.

Step II: Start with a 1000 Hz tone at a level above the threshold to allow easy identification of the tone. This tone is selected because it is an important speech frequency, and the patient is less apt to mistake the frequency. If the patient is suspected to be having a profound hearing loss then the testing should be started with 250Hz frequency. This is because of the fact that the individuals with profound hearing loss often have testable hearing only in the low frequency range.

Step III : The patient's understanding of the listening task should be checked by using both short and long duration test tones. The patient should be instructed to raise the index finger as soon as the sound is heard.

Step IV : During testing, the examiner should vary the interval between tone presentations to avoid telegraphing the stimulus. Tone should not be presented while the attenuator dial is being rotated, because switching artefacts may contaminate the results. As the threshold levels are being reached, a check should be made for the existence of abnormal tone decay. This is done by sustaining the tone for several seconds longer than usual. If the index finger drops before the tone is discontinued, abnormal tone decay should be suspected.

Step V: The starting intensity of the test tone is reduced in 10 dB steps following each positive response, until a hearing threshold level is reached at which the subject fails to respond. Then, the tone is raised by 5 dB, if the subject hears this increment, the tone is reduced by 10 dB; if the tone is not heard then it is raised by another 5 dB increment. This 5 dB increment is always used if the preceding tone is not heard, and a 10 dB decrement is always used when the sound is heard. The threshold is defined as the faintest tone that can be heard 50% or more of the time, and is established after several threshold crossings.

If there is no response at the maximum output of the audiometer, an arrow pointing downward should be attached to the symbol designating the test ear and placed on the audiogram at the hearing threshold level coinciding with the maximum output for the test frequency. If the tone is heard at the minimum level, the audiogram should be marked in similar fashion but should have the arrow pointing upward.

Step VII: Testing of the second ear should begin with the last frequency used to test the first ear. There is no need to start again with a 1000 Hz tone because if one side of the heard has learned the listening task, the other side knows it as well. The test is terminated after all desired frequencies have been examined.

Interpretation: With air conduction readings alone one cannot reliably come to a diagnosis. Even a basic distinction between conductive and sensorineural hearing loss cannot be made with air conduction hearing tests. When there is a hearing loss in air conduction audiogram, it represents cumulative deficits from the outer, middle, inner ear and retro cochlear system of a subject. Bone conduction audiometry is a must before a classification of the deafness could be made.

Scale of hearing threshold estimation in air conduction audiometry:

dB loss	Description
-10 – 15	Normal limits
16-19	Mild hearing loss
30-44	Moderate hearing loss
45-59	Moderately severe hearing loss
60-79	Severe hearing loss
80 and above	Profound hearing loss



Figure showing bone vibrator

In bone conduction audiometry high frequencies cannot be used for testing. Frequencies above 4000 Hz cannot be used because they are beyond the vibrating capabilities of the bone vibrator.

Citation:

- 1.

Pure tone audiometry by drtbalu [Internet]. [cited 2012 Jan 1];Available from:
http://drtbalu.co.in/pt_audio.html

Congenital anomalies of pinna and external auditory canal

Anomalies of Pinna and external auditory canal can be termed as microtia and congenital aural atresia respectively.

Definition:

Microtia: is defined as the abnormal development of the Pinna resulting in a malformed auricle. The deformities caused could range from mild distortion of the anatomic landmarks to the complete absence of auricle.

Congenital aural atresia: is defined as a failure of development of external auditory canal. Congenital aural atresia is always associated with a certain degree of microtia.

Microtia and congenital aural atresia are rare deformities. These deformities are always associated with other malformations of auditory system.

Etiology:

1. Exposure to teratogens like vitamin A
2. Vascular insults and genetic aberrations
3. Isolated microtia can occur with branchial arch anomalies
4. Can occur as a part of a single gene deletion or embryopathic development anomalies, eg. Goldenhar syndrome
5. Certain auricular deformities can occur as a result of multifactorial insults to the developing foetus

Incidence: Microtia and congenital auditory atresia occur in approximately 1 in every 20,000 live births. These deformities commonly occur unilaterally, more so, on the right side. Men are affected thrice as common as women. The degree of auricular deformity usually correlates with the degree of middle ear deformity. The incidence of inner ear deformities is very rare in patients with congenital auditory atresia. Microtia is associated with other anomalies of face 50% of the time.

Women with four or more pregnancies are at increased risk of bearing a child with microtia. The incidence of microtia is higher in Japanese population.

Embryology: During the sixty week of intrauterine life the external ear begins to develop around the dorsal end of the first branchial cleft. On either side of this cleft lie the first (mandibular) and second (hyoid) arches. The auricle develops from these arches as 6 small buds of mesenchyme known as the six hillocks of His. The first arch gives rise to the hillocks

1 to 3 and the second arch gives rise to hillocks 4 to 6.

Traditional theory suggests that hillock 1 becomes the tragus, hillocks 2 and 3 from the helix, hillocks 4 and 5 form antihelix and hillock 6 forms the lobule of the ear. More recently it has been suggested that the second arch contributes approximately to 85% of the auricle. The lobule is the last component of the pinna to develop.

The auricle begins to develop in the anterior neck region, then it is postulated to migrate dorsally and cephalad as the mandible begins to develop during the second and third months of gestation. By the 5th month of gestation the pinna lies in its adult location.

The external auditory canal begins to develop from the first branchial cleft during the first two months of gestation. During the first month a solid epithelial cell rest forms in this area and is in contact with the endoderm of the first pharyngeal pouch. There is an intervening mesoderm preventing direct contact between the ectoderm and endoderm.

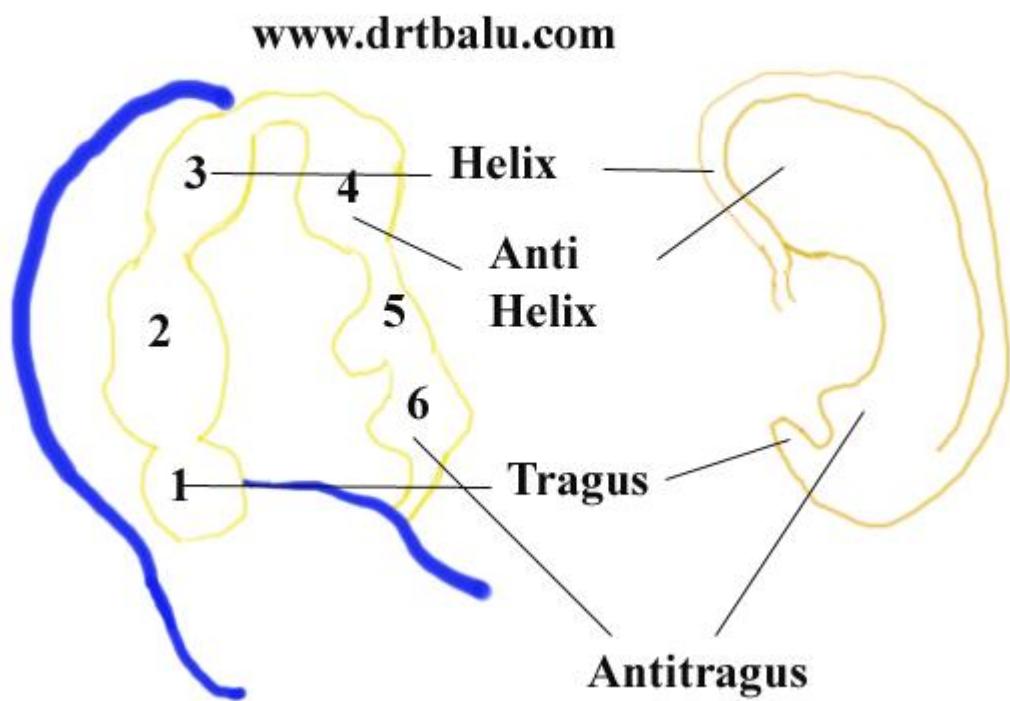


Illustration showing embryology of pinna

Applied anatomy of pinna: Pinna reaches mature size between ages of 13 - 15. The superior edge of the pinna should be in line with the lateral edge of eyebrow or upper eyelid. It also shows a posterior inclination ranging from between 5 - 30 degrees. The angle of the ear is parallel to that of the dorsum of nose, usually within a range of 15 degrees.

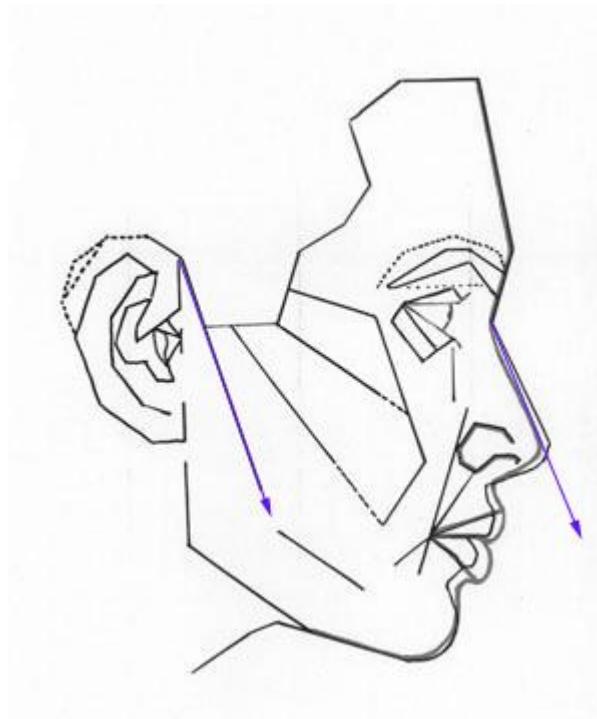


Diagram showing the angle of the ear running parallel to that of dorsum of the nose

Evaluation:

While examining a patient with microtia attention should be paid to the mandible, oral cavity, cervical spine and eyes. This is done to rule out other associated cervico facial congenital anomalies. The quality of skin over the malformed pinna should also be noted. The integrity of facial nerve should be tested and documented.

Patients with unilateral microtia and congenital aural atresia usually have normal hearing on the opposite ear. Hearing status on the affected side should be recorded.

The role of CT scan in these patients is to assess the middle ear anatomy and inner ear anatomy. The presence of congenital cholesteatoma is common in these patients and hence must be sought in the CT scan of the affected side.

In order to select ideal candidates for repair microtia has been classified by Marx as:

Grade I : The pinna is malformed and smaller than normal. Most of the characteristics of the pinna, such as the helix, triangular fossa, and scaphae, are present with relatively good definition

Grade II: The pinna is smaller and less developed than in grade I. The helix may not be fully developed. The triangular fossa, scaphae, and antihelix have much less definition

Grade III: The pinna is essentially absent, except for a vertical sausage-shaped skin remnant. The superior aspect of this sausage-shaped skin remnant consists of underlying unorganized cartilage, and the inferior aspect of this remnant consists of a relatively well-formed lobule

Grade IV: Is complete anotia.

Two other classification systems one by Jahrsdorfer and the other by De la cruz are commonly used these days. De la cruz classification divides the malformations into major and minor categories. Obviously ears with minor deformities are better surgical candidates for successful reconstruction, whereas ears with major deformities should be managed with hearing aids.

Surgical therapy for unilateral microtia: This is performed as a staged procedure using autogenous rib cartilage. This technique was refined by Tanzer.



Figure showing microtia of pinna and atresia of external auditory canal

Tanzer surgery: Is performed in four stages. There is a three month gap between these stages.

First stage: Rib cartilage is harvested and sculptured into the shape of pinna and is placed under skin pocket of the microtic ear.

Second stage: Formation of the lobule

Third stage: Elevation of the ear with insertion of a post auricular skin graft

Fourth stage: Formation of the tragus with a skin/cartilage composite graft from the contralateral ear and full-thickness skin graft for the conchal area from the contralateral ear

Nagata technique: This is a two staged procedure developed by Nagata. This involves constructing the auricular framework from the sixth to ninth rib cartilages. The framework is created using stainless steel sutures. The framework is placed and the lobule remnant is transposed. Six months later a reconstruction is performed and the graft is released.

The aim of congenital aural atresia repair is to provide serviceable hearing to the patient. Children associated with congenital aural atresia with other syndromes like Treacher Collins syndrome should not be operated upon, and should be managed with implantable hearing devices. Congenital aural atresia should be performed two months after microtia repair to preserve blood supply to skin and subcutaneous blood supply.

Before proceeding with repair of congenital aural atresia a high resolution CT scan must be performed. The mastoid cavity and middle ear anatomy should be completely analysed. The presence of a mastoid cavity is a must for surgery because the canal is created at the expense of the mastoid cavity.

The factors that must be taken into consideration before performing the surgery include:

1. Status of inner ear
2. Temporal bone pneumatisation
3. Course of facial nerve
4. Presence of foot plate and round window
5. Presence of cholesteatoma

Three possible approaches can be followed for congenital aural atresia repair. They are:

1. Mastoid
2. Anterior
3. Modified anterior

Mastoid approach: In this approach the external auditory canal is created at the expense of mastoid cavity. It involves drilling out the mastoid and identifying the sino dural angle. This is a risky procedure because of distorted anatomy of the facial nerve in these patients.

Anterior approach: Is the most common approach used these days. In this approach a post auricular incision is made and the subcutaneous tissue and periosteum are raised anteriorly up to the level of glenoid fossa. If any remnant of tympanic bone is present drilling is started at the cribriform area, and if no tympanic bone is present the drilling begins at the temporal line just posterior to the glenoid fossa. Drilling is continued anteriorly and

medially till epitympanum is entered. The most common anomaly encountered in the middle ear of these patients is a fused malleal - incudal joint. Stapes is usually normal in these patients. The atretic bone is carefully removed uncovering the ossicles. The facial nerve usually lies medial to the ossicular mass, and must be protected at all costs. It could commonly be injured in the posterior - inferior middle ear space. Drilling is continued till the canal is about 10mm in size. Ossicular chain reconstruction is performed and a neo tympanum is fashioned using temporalis fascia graft. Split thickness skin graft is used to line the external auditory canal. A wide meatoplasty is fashioned and a large wick is inserted to stent the canal.

Modified anterior approach: This approach is used in patients with a thick atretic plate because of poor orientation during dissection. This poor orientation may risk carotid artery, facial nerve, and lateral semicircular canal to injury. Orientation in these patients could be achieved by an initial posterior dissection up to the level of sinodural angle. This enables the surgeon to identify the level of lateral canal and ossicular mass. From here on the approach is similar to that described under anterior approach.

Complications of these surgical procedures:

1. Injury to facial nerve
2. Injury to lateral canal
3. Sensorineural hearing loss
4. Rejection of graft material
5. Restenosis
6. The morbidity associated with rib harvest is significant and includes scarring, deformity and risk of pneumothorax.

Prosthetic devices: Tissue expanders and other prosthetic materials can be used in these surgical procedures. The use of tissue expanders before implantation of microtia framework can avoid the use of skin grafts and could also reduce the number of surgical procedures.



Father of otoplasty Tanzer

Citation:

1. Congenital anomalies of pinna and external auditory canal by drtbalu [Internet]. [cited 2012 Jan 1]; Available from: <http://drtbalu.co.in/microtia.html>

BERA

Synonyms: Brain stem evoked response audiometry, Auditory brain stem response, ABR audiometry, BAER (Brainstem auditory evoked response audiometry).

Definition: Bera is an objective way of eliciting brain stem potentials in response to audiological click stimuli. These waves are recorded by electrodes placed over the scalp. This investigation was first described by Jewett and Williston in 1971.

Even though BERA provides information regarding auditory function and sensitivity, it is not a substitute for other methods of audiological evaluation. It should be always viewed in conjunction with other audiological investigations.

Procedure: The stimulus either in the form of click or tone pip is transmitted to the ear via a transducer placed in the insert ear phone or head phone. The wave forms of impulses generated at the level of brain stem are recorded by the placement of electrodes over the scalp.

Electrode placement: Since the electrodes should be placed over the head, the hair must be oil free. The patient should be instructed to have shampoo bath before coming for investigation. The standard electrode configuration for BERA involves placing a non-inverting electrode over the vertex of the head, and inverting electrodes placed over the ear lobe or mastoid prominence. One more earthing electrode is placed over the forehead. This earthing electrode is important for proper functioning of preamplifier.



Figure showing BERA equipment

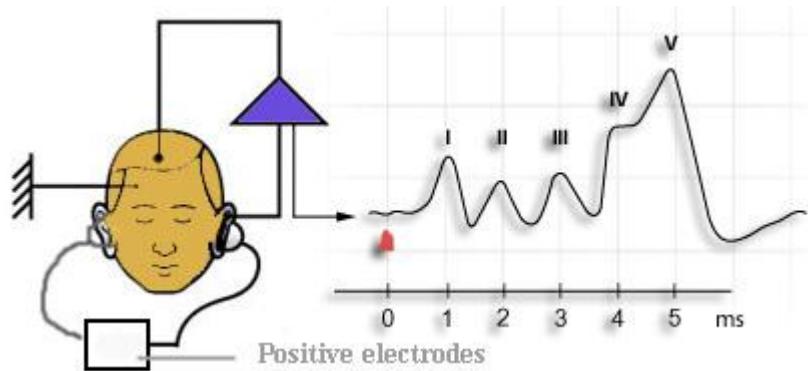


Figure showing placement of BERA electrodes

Since the potentials recorded are in far field, well displaced from the site of impulse generation, the wave forms recorded are very weak and they need to be amplified. This amplification is achieved by improving the signal: noise ratio.

How to improve signal to noise ratio: Three parallel approaches are designed to achieve this goal.

Filtering: This is employed to reduce the recording bandwidth so that only the important components of the signal generated are recorded.

Repeated stimulation: This is done with synchronous time domain averaging to increase the amplitude of the components of the signal. In real time situations these two can be achieved by connecting the recording electrodes to a preamplifier, with appropriate filter settings.

Polarity alteration: By altering the polarity of impulses recorded, the artefacts are cancelled making the brain stem waves stand out.

In auditory brain stem evoked response audiometry, the impulses are generated by the brain stem. These impulses when recorded contain a series of peaks and troughs. The positive peaks (vertex positive) are referred to by the Roman numerals I - VII.

These peaks are considered to originate from the following anatomical sites:

1. Cochlear nerves - waves I and II
2. Cochlear nucleus - wave III
3. Superior olivary complex - wave IV
4. Nuclei of lateral lemniscus - wave V
5. Inferior colliculus - waves VI and VII

These peaks occur in most readable form in response to click stimuli over a period of 1 - 10 milliseconds after the stimulus in normal hearing adults.

BERA is resistant to the effects of sleep, sedation, sleep and anesthesia. Its threshold has been found to be within 10dB as elicited by conventional audiology.

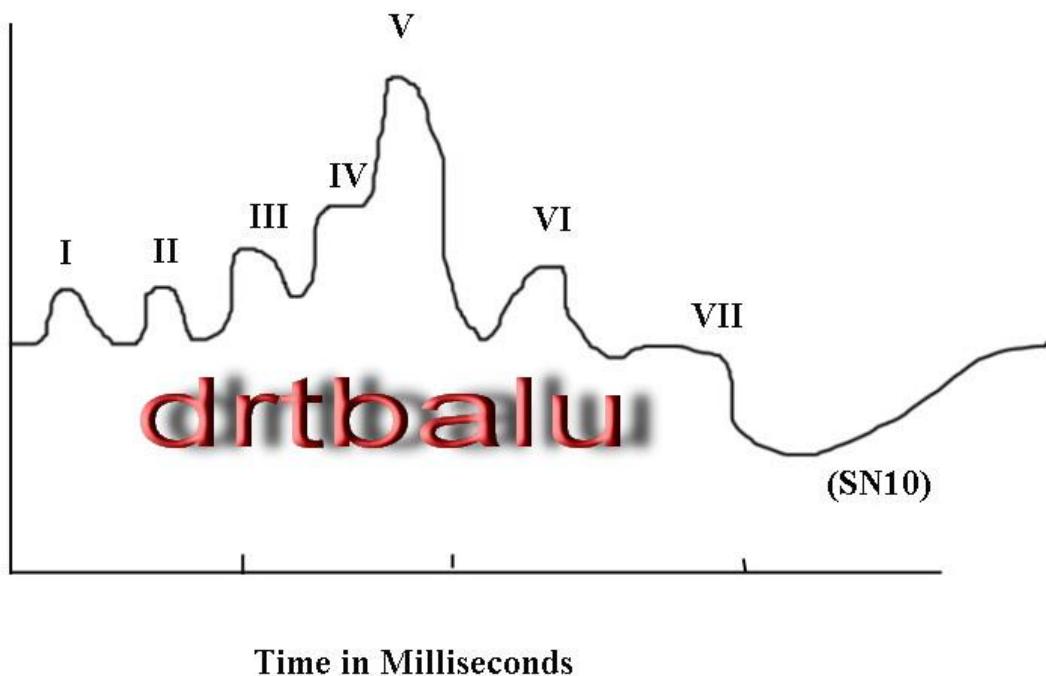


Illustration showing various wave forms of BERA

There are two different types of auditory evoked potential tests. These tests are being used as an adjunct to routine diagnostic testing. The two types of auditory evoked potential tests are: 1. Auditory brain stem response and 2. Auditory cortical response.

Auditory cortical response: Records the impulses generated by brain in response to tone stimuli. It is recorded using cortical response audiometry (CERA). CERA is very useful for threshold estimation of hearing, whereas BERA is highly useful for objective threshold estimation of hearing as well as differential diagnostic purposes. These responses are more generalised and originate from the brain cortex occurring between 50 - 300 milliseconds after the onset of stimulation. Since these responses are generally elicited with a tone burst

lasting approximately for about 200 milliseconds, its responses are highly frequency specific. This is in contrast to BERA because brain stem responses are evoked by click stimuli and are not frequency specific. Interpretation of CERA is easy and straightforward. Threshold is defined as the minimum stimulus level that gives a consistent and identifiable response. The patient must be lying still during the recording process. This test is hence unsuitable for young children who may not co-operate.

Differences between BERA and CERA:

BERA	CERA
Recording is made from brain stem potentials	Recording is made from cortical potentials
Click stimulus is used	Tone stimulus is used
Responses are not frequency specific	Responses are frequency specific
Can be performed in awake and restless patients	Patient should lie still during the entire process
Responses begin after 1 - 10 milliseconds after stimuli	Response begins after 50 - 300 milliseconds after stimulation
Suitable for even young children	Unsuitable for children

Uses of BERA:

1. It is an effective screening tool for evaluating cases of deafness due to retrocochlear pathology i.e. (Acoustic schwannoma). An abnormal BERA is an indication for MRI scan.
2. Used in screening newborns for deafness
3. Used for intraoperative monitoring of central and peripheral nervous system
4. Monitoring patients in intensive care units
5. Diagnosing suspected demyelinated disorders

BERA findings suggestive of retrocochlear pathology:

1. Latency differences between interaural wave 5 (prolonged in cases of retrocochlear pathology)
2. Waves I - V interaural latency differences - prolonged
3. Absolute latency of wave V - prolonged

4. Absence of brain stem response in the affected ear

BERA has 90% sensitivity and 80% specificity in identifying cases of acoustic schwannoma. The sensitivity increases in proportion to the size of the tumor.

Criteria for screening new-born babies using BERA:

1. Parental concern about hearing levels in their child
2. Family history of hearing loss
3. Pre and post natal infections
4. Low birth weight babies
5. Hyperbilirubinemia
6. Cranio facial deformities
7. Head injury
8. Persistent otitis media
9. Exposure to ototoxic drugs

Citation:

1.
BERA (Brainstem evoked response audiometry) by drtbalu [Internet]. [cited 2012 Jan 1];Available from: <http://drtbalu.co.in/bera.html>

Otoacoustic emissions

Definition: Otoacoustic emissions (OAE) are sounds produced by motile elements of cochlear outer hair cells. These sounds can be recorded easily. These sounds were first identified and reported by Kemp in 1978.

The main function of OAE tests is to assess the function of cochlear hair cells. These tests could be used:

1. To screen children and neonates for hearing disabilities
2. Estimate hearing sensitivity within a limited range of frequencies
3. To differentiate sensory and neural components in sensorineural hearing loss
4. To rule out malingering (functional hearing loss)



Image showing OAE equipment

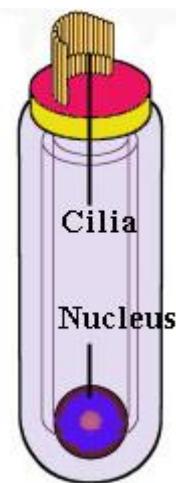


Image showing outer hair cell

Since the measurement of otoacoustic emissions are recorded from the external auditory canal, the integrity of the middle ear and the cochlear function come into play.

Role played by otoacoustic emissions: As explained earlier otoacoustic emissions are generated by the outer hair cells of cochlea. These otoacoustic emissions play the role of cochlear amplifier. In sensori neural hearing loss the cochlear amplification is lost leading to:

- a. Reduction in the hearing level
- b. Reduction in the clarity of spoken words

Types of Otoacoustic emissions: OAE's are classified according to the stimulus employed to elicit them, or by the mechanism that causes them.

Spontaneous otoacoustic emission: These narrow band continuous signals occur without any stimulus. These signals can be detected in a majority of persons with normal pure tone threshold. The clinical value of this signal is limited, as this is not present in all normal ears. The absence of spontaneous otoacoustic emission does not imply cochlear dysfunction.

Synchronised otoacoustic emissions: are potentials generated by outer hair cells of cochlea which are synchronised to the external stimuli using time averaging techniques.

Measurement of these potentials is difficult and highly cumbersome. They do not have any diagnostic / prognostic value as it is not a consistent feature in all normal ears.

Three types of OAE's have been recorded in response to various stimuli. These are also known as evoked oto acoustic emissions.

Stimulus frequency otoacoustic emission: These potentials are evoked with some kind of acoustic stimulus. The evoking acoustic stimulus is a pure tone one with a low intensity

level. Study of these potentials are still in the experimental stage, and hence not widely used.

Transient otoacoustic emission: (TOAES) These potentials are also known as transient evoked otoacoustic emission. Since it was first described by Kemp it is also known as Kemp echoes. The time delay between the stimulus and response allows the examiner to isolate these responses. These echoes recorded from normal ears always mirror the spectrum of the stimulating sound impulse. The probe used to record transient otoacoustic emission has two openings, one for the presentation of a single stimulus like a click, and the other opening which is used to record the transient evoked otoacoustic emission. The second opening is connected to a microphone to enable recording to take place.

Clicks are commonly used as stimuli, sometimes tone burst stimuli can also be used. The stimulus used should be 80 - 85dB sound pressure level. The rate of stimuli should be at least 60 / minute. When present TOAES occur at frequencies of 500 - 4000 Hz

Distortion Production Otoacoustic emission: (DPOAEs): These are low intensity signals that occur during stimulation of the ear. A common way to record them is to present the ear with two continuous signals called the primary tones and analyse the spectrum of sounds detected at the external auditory canal. The intensity levels of the signals used are 55 dB and 65 dB respectively.

Prerequisites for obtaining otoacoustic emissions:

1. Unobstructed external auditory canal
2. Perfect seal of external auditory canal with the probe
3. Optimal positioning of the probe
4. Absence of middle ear pathology
5. Functioning cochlear outer hair cells
6. A relatively still patient
7. Quiet recording environment

Interpretation of the recordings made:

Spontaneous otoacoustic emissions: are commonly found in 50% of individuals with normal hearing. It is generally not seen in patients with less than 30 dB hearing level. If spontaneous otoacoustic emissions are elicited in a patient the cochlea could be assumed to be in good

health. These emissions are more bilateral than unilateral. They are more commonly recorded in females than in males.

These spontaneous otoacoustic emissions are not associated with tinnitus because the associated cochlear abnormality causes the SOAEs to disappear.

Transient otoacoustic emissions: are usually used to screen neonates for hearing disabilities. These impulses can be recorded only in response to short and transient stimuli. These impulses have very limited frequency specificity. The presence of these emissions suggests the cochlear sensitivity in the region of 20 - 40 dB or better.

Distortion product otoacoustic emissions: These emissions have greater frequency specificity. These potentials are useful in detection of early detection of cochlear damage i.e. due to noise or drug exposure.

Causes of absent otoacoustic emissions:

Non pathological:

1. Poor probe tip placement
2. Standing waves
3. Cerumen occlusion
4. Vernix caseosa in infants
5. Unco operative patient

Pathologic causes:

Outer ear: Stenosis, otitis externa, cysts etc.

Tympanic membrane: Perforations. Grommets usually don't complicate recordings

Middle ear: Otosclerosis, ossicular disruption, cholesteatoma, otitis media

Cochlea: Exposure to ototoxic drugs, Noise exposure

Central auditory disorders don't affect otoacoustic emissions.

Conclusion:

Otoacoustic emissions play an important role in screening infants for disorders of hearing.

Citation:

1.
Otoacoustic emission by drtbalu [Internet]. [cited 2012 Jan 1];Available from:
http://drtbalu.co.in/oto_emission.html

Endolymph

Definition:

Endolymph is the fluid that fills the scala media of the cochlea. Hence this space is also known as endolymphatic space / Scarpa's space.

The walls surrounding the endolymphatic space are said to have occluding tight junctions between the cells obstruction ionic movements in and out of this compartment. Commonly accepted borders of this space are shown in the figure above.

This endolymphatic space extends throughout the membranous labyrinth and is joined to the endolymphatic sac by the endolymphatic duct.

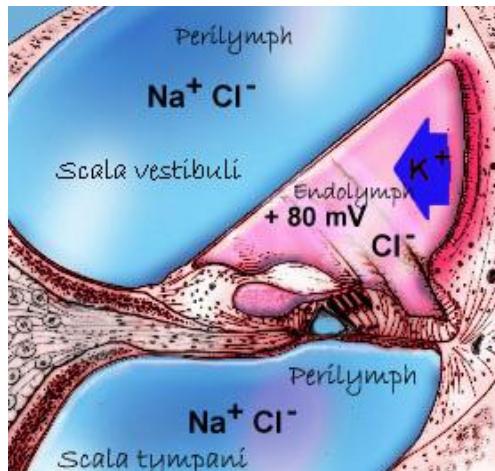


Figure showing inner ear compartments

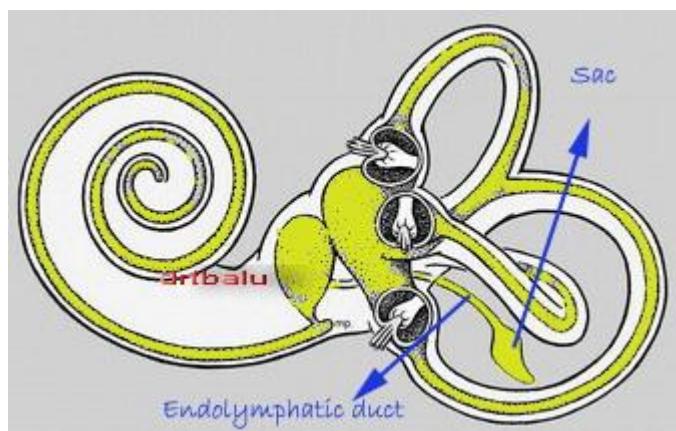


Figure showing vestibule of inner ear

Site of secretion of endolymph:

It has generally been accepted that endolymph of the cochlea is produced by stria vascularis.

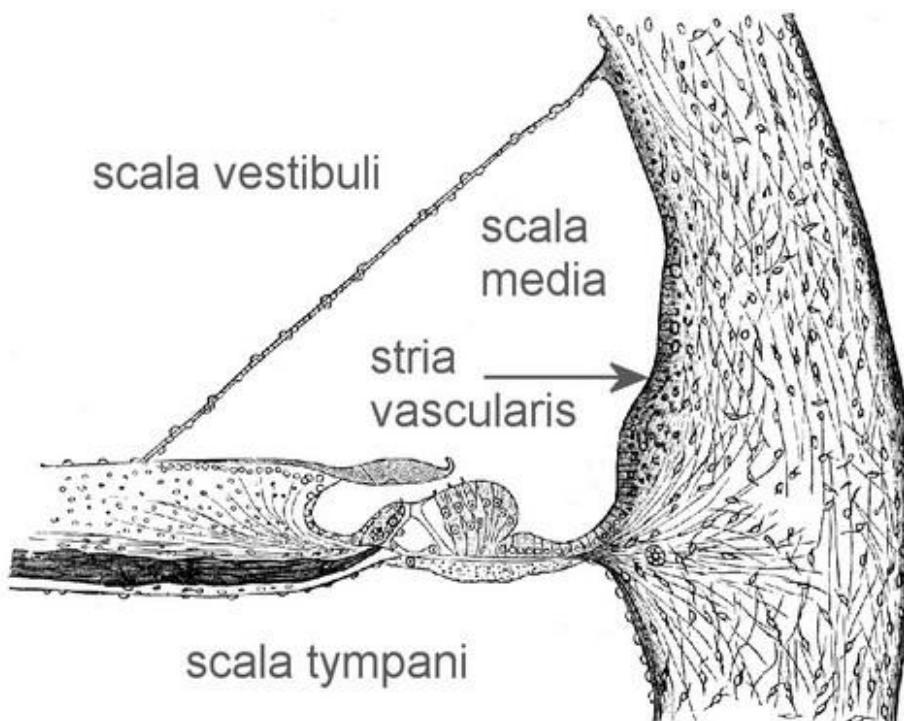


Image showing stria vascularis

The cells of stria vascularis are secreting cells. Under light microscope this area can be divided into:

1. Superficial - Dark stained cells (marginal cells)
2. Basal cells - Lightly stained cells

When seen under electron microscope the marginal cells show long infoldings on its basal edge i.e. the edge farthest from the endolymph. These infoldings contain numerous mitochondria. It is here that energy consuming pumping takes place in the stria vascularis.

Composition of endolymph:

Endolymph is very unique among extracellular fluids in that it contains high concentrations of potassium and low concentrations of sodium resembling more the intracellular fluid composition.

Concentration of potassium in endolymph varied from 144 - 188 mM.

Concentration of sodium in endolymph varied from 0.2 - 2 mM.

The endolymph has a positive charge ranging from + 50 - +120 mV. The potentials can still be larger close to stria vascularis. This positive charge is maintained by the activity of energy consuming $\text{Na}^+ + \text{K}^+$ -ATPase in the marginal cells of stria vascularis. This potential is also known as endocochlear potential.

How this endocochlear is potential generated?

The current accepted model of endolymphatic fluid potential generation is the one proposed by Wangemann and Marcus et al. This model in fact relies on the complex geometrical orientation of striae spaces.

The steps include:

1. Removal of potassium from the intrastrial space by active pumping in the basal membranes of marginal cells.
2. This reduces the level of potassium in the intrastrial space
3. Diffusion of potassium occurs through the potassium channels between the intermediate cells and the intrastrial space.
4. This generates a diffusion potential between the intrastrial space and the intermediate cells
5. Intrastrial space becomes highly positive thus giving rise to endocochlear potential.

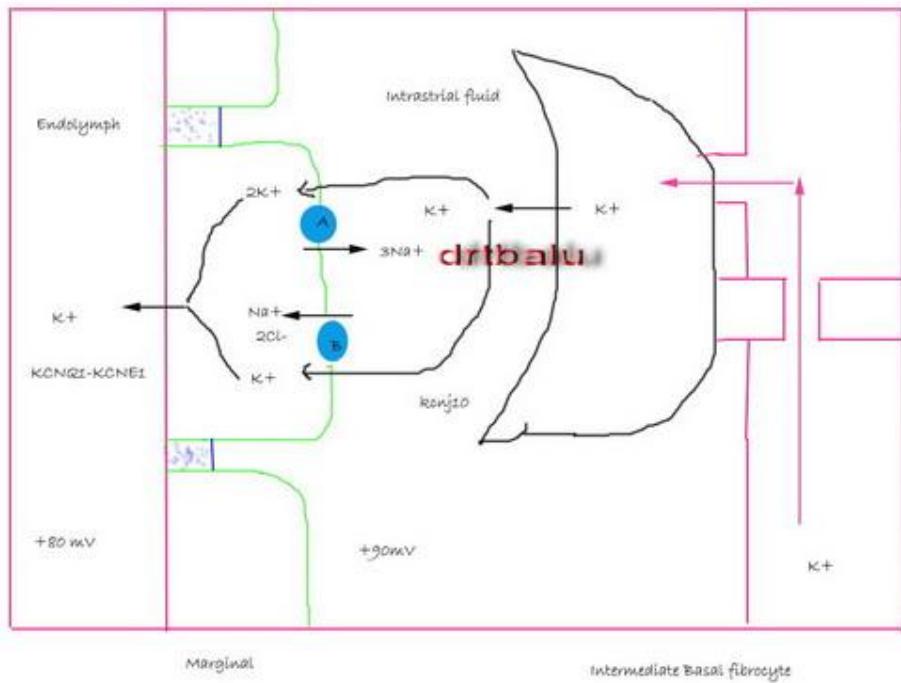


Figure showing Wangemann theory

Citation:

1. Endolymph by drtblu [Internet]. [cited 2012 Jan 1]; Available from: <http://drtblu.co.in/endolymph.html>

Electrocochleography

Definition: Electrocochleography is a procedure to record the potentials generated by the cochlea and the auditory nerve. There are three classes of potentials that can be recorded from the cochlea and auditory nerve in response to a sound stimuli. They are compound action potential of auditory nerve (AP), Summating potential (SP), and cochlear potential (CP) also known as cochlear microphonic.

This is an objective way of estimating hearing threshold of a patient. When these recordings are made from electrodes placed close to the source of impulse the recorded potentials are quite large. If the recording is made from electrodes placed in far field, the potentials recorded are small in nature. It is always advisable to place the recording electrodes as close to the source of impulse as possible i.e. over the promontory.

Types of electrodes: Three types of electrodes are used to record ECog. They are:

Transtympanic membrane electrodes: These are needle electrodes, which are placed over the promontory by penetrating the ear drum. The ear drum must be anesthetised before placing the electrode. The needle passes through posterior inferior quadrant of ear drum. These electrodes are 22 gauge hypodermic needles.

Intrameatal electrodes: These electrodes are placed in the external auditory canal.

Surface electrodes: These are attached outside the external canal. These electrodes are clipped to the earlobe.

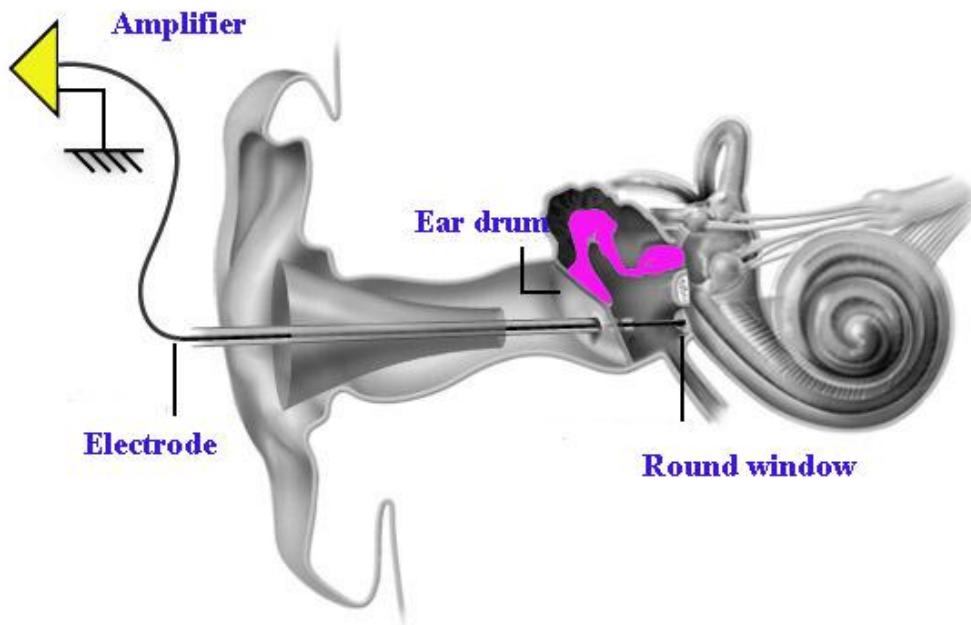


Figure showing electrode placement in Electrocochleography

Advantages and disadvantages of transtympanic electrode:

Recordings from the promontory reveal evoked response, whose amplitudes are larger than that obtained by placing electrodes in the far field, like external canal or pinna. This is the advantage of using a transtympanic electrode in recording ECog. In fact this is the only advantage of using this electrode. The whole procedure is invasive, with the attendant complications of anesthetic medication.

Advantages and disadvantages of intrameatal and surface electrodes:

Since recordings from these electrodes are far field, the impulses recorded are of low amplitudes, and need to be augmented. The main advantage of these electrodes is that they are non-invasive, and patient need not be anesthetised before placing the electrode. These electrodes can be used in screening procedure in neonates.

Type of acoustic signal used to stimulate Ecog: The appropriate signal used to stimulate ECog should be abrupt so that a large number of acoustic nerve fibers are stimulated. Click signals are the most appropriate stimuli. Tone pips can also be used. The sound pressure levels used should be between 80 - 85 dB.

Action potential: This is an A.C. potential representing the summed potential of synchronous firing of thousands of auditory nerve fibers. Action potentials are dominated by contributions from high frequency nerve fibers as a response to transient stimuli of rapid onset. It occurs at the onset of stimulus. It can be produced by clicks or tone pips. It is characterised by predominantly negative peaks i.e. N1 and N2. These waves are independent of stimulus phase and duration.

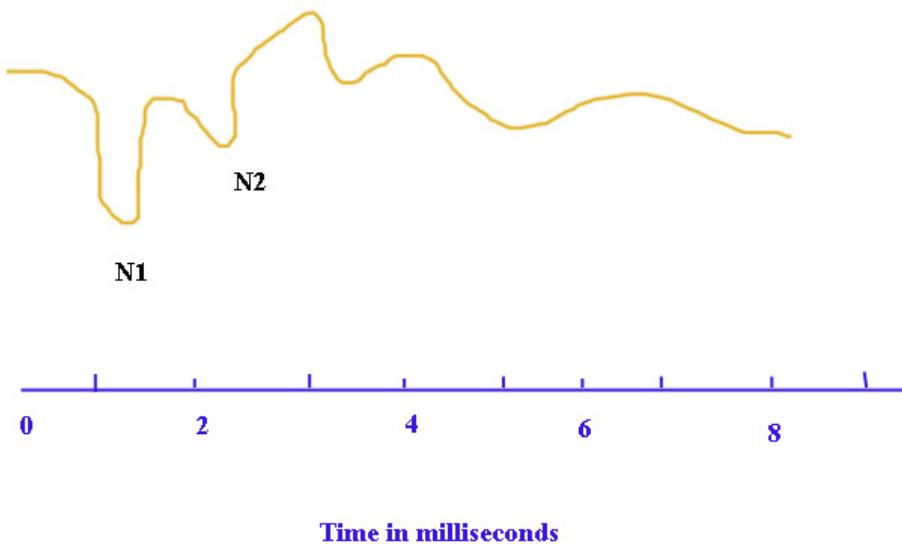


Figure showing action potential recording

The most useful features of action potential curve are its magnitude and latency. Out of them the magnitude has a direct relationship to the number of nerve fibers firing. Latency represents the time interval between the onset of stimulus and the peak of N1. The presence of action potential is dependent on the proper functioning of acoustic nerve fibers. Wave N1 is absent in patients with retrocochlear lesions.

Cochlear microphonics: These are generated predominantly by the outer hair cells of cochlea. This is an A.C. potential. The basal turn of cochlea play an important role in generation of cochlear microphonics. This is more so when the recording is made from electrodes close to the round window. Its phase and duration are mostly stimulus dependent. It is diminished when stimulus is present with alternating polarity. It helps in differentiating cochlear from nerve deafness.

Summating potential: This is direct current. Outer hair cells of cochlea are responsible for this potential. The direction of this potential is dependent on complex interaction between the stimulus parameters and the location of the recording electrode.

Uses:

Electrocotchleography is predominantly used in the diagnosis, assessment and monitoring Meniere's disease, and endolymphatic hydrops. In Meniere's disease the summation potential is enlarged. It is also used for intra operative monitoring of peripheral auditory system. It is also useful in objectively assessing auditory thresholds.

Citation:

1.
Electrocotchleography by drtbalu [Internet]. [cited 2012 Jan 1];Available from:
http://drtbalu.co.in/ecoc_g.html

Otophyma

Introduction:

Otophyma is the term used to describe sebaceous gland hypertrophy and hyperplasia of the pinna. It is usually considered to be the end stage of rosacea. This disorder is considered to be a chronic cutaneous disorder of unknown etiology.

Phyma is a greek word meaning “growth”. Usually it is caused by sebaceous gland hyperplasia and hypertrophy of the surrounding fibrous tissue. These Phymas are named according to the site of affliction.

Rhinophyma – when nose is affected.

Gnatophyma – When chin is affected

Metophyma – when forehead is involved

Otophyma – When pinna is involved

Blepharophyma – When eyelids are involved

Otophyma or rosaceous lymphoedema can either be unilateral / bilateral. Even though Rosacea is a common cutaneous disorder of unknown etiology affecting females otophyma is very rare.

Diagnosis:

The diagnosis of otophyma is made purely on clinical basis. Biopsy is necessary to differentiate this from similar looking diseases like lupus, carcinoma and angiosarcoma. Histopathology reveals hyperplasia and hypertrophy of sebaceous glands. The ducts of these glands appear dilated, convoluted and plugged. There is also associated proliferation of fibroblasts and presence of bacteria and inflammatory cells. Demodex folliculorum mite is commonly seen.

Management:

Early lesions which happen to be hyperaemic respond to oral antibiotics like tetracycline and metronidazole. Late stages may require surgery.

Surgery:

1. Full thickness excision with split thickness graft
2. Full thickness excision with full thickness graft
3. Decortication – partial excision / shaving of the lesion. Cryo surgery has also been used in decortication procedures

SISI Test

Introduction: The SISI test is still widely used to determine whether the patient is having cochlear pathology. This test is based on a phenomenon known as recruitment (abnormal loudness growth). It was Dix and Hallpike who related the presence of loudness recruitment to cochlear pathology.

Difference limen for intensity (DLI): is the smallest change in the intensity of a pure tone which can just be detected. It is usual for patients with normal hearing to have difficulty in detecting small changes in intensity close to threshold. Patients with cochlear pathology will be able to appreciate the change in intensity better because of the phenomenon of recruitment. DIL could safely be assumed to be an indirect indicator of the phenomenon of recruitment. It was Luscher and Zwischenlocki who developed a DIL test which remained popular for quite sometime.

SISI test proper was introduced by Jerger and Shedd in 1959 as a test for the phenomenon of recruitment. In this test a pure tone was presented to the patient at a sound level of 20dB. A small increase in intensity is superimposed upon the steady state tone at periodic intervals. The size of the increment varied from 5 - 1 dB. Jerger clearly demonstrated that the ability to detect the 1 dB increments was restricted to patients with cochlear pathology. This ability was absent in patients with normal hearing or with a conductive hearing loss. This test is very simple to perform and is less confusing than the DIL test. The SISI test differs from the classical DLI tests in that the patient's precise DLI is not explored. It just tests the cochlea's ability to respond to a transient signal of small amplitude.

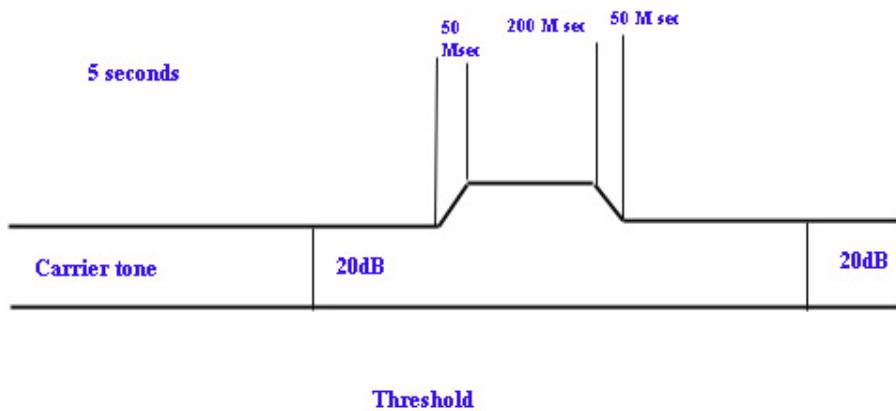


Diagram showing the signal used to perform SISI

As shown in the fig. a carrier tone is introduced into the patient's ear at a Sound level of 20 dB. Every 5 seconds a short increment is superimposed, starting with 5 dB increments. The signal has an on-off time of 50 msec and 5 seconds elapse between increments. The patient is instructed to indicate when he hears a brief jump in the loudness of the tone. After 5 such jumps (to condition the patient) the size of the increment is lowered to 1 dB marking the beginning of the SISI test. Twenty 1 dB increments are introduced and the subject is required to indicate when the increment is heard each time. If a number of consecutive increments heard exceeds 5 then the examiner should delete several increments to ensure that the subject is responding to the change in intensity and not the time interval. If the patient fails to respond to several increments in a row the increment size can be increased for retraining the patient before proceeding with the test proper. These steps will avoid false negative and false positive results.

SISI test variants:

There are 5 different variants of SISI tests.

1. One dB increments at 20dB sound level (classical SISI) - High scores suggest a cochlear lesion.
2. Two to 5 dB increments at 20 dB sound level - Low scores suggest a retrocochlear lesion.
3. One dB increments at high sound levels (75dB) - Low scores suggest a retrocochlear lesion.
4. Increment sizes varied from 1 - 5 dB at 20 dB sound level - poorer scores in one ear than the other (when their thresholds are approximately equal) suggests a central lesion opposite the ear with the lower score.
5. One dB increments at sound levels ranging from 20 dB to high levels (about 75dB) in 10 dB steps for both ears. Difference in the rate at which scores increase suggests a retrocochlear lesion. The disorder is located on the same side as the ear which has not shown normal increases in intensity.

Uses of SISI test:

This test helps in determining the site of lesion in the auditory system by determining whether a disorder is cochlear or noncochlear. The diagnostic accuracy of this test depends upon the amount of hearing loss a patient may have.

Citation:

1.
SISI test by drtbalu [Internet]. [cited 2012 Jan 1];Available from: <http://drtbalu.co.in/sisi.html>

Gradenigo syndrome

Introduction:

This syndrome was first described by Gradenigo in 1907. He was an Italian Otologist. Maurice Lannois added vital data to those already described by Gradenigo. Gradenigo classically described the following as the classic features of this syndrome:

1. Discharging ear
2. Retro orbital pain
3. Adbucent nerve paralysis causing diplopia

Causative factors:

1. Uncontrolled mastoiditis
2. Epidural abscess following mastoiditis

Pathophysiology:

This syndrome has been known to occur due to spread of ear infection to involve air cells around petrous apex. It is hence also known as "Petrosus apex syndrome" / "Petrosus apicitis". Infection & inflammation of petrous apex involves 6th cranial nerve at the Dorello's canal and 5th cranial nerve in the Meckel's cave. Meckel's cave lies close to Dorello's canal. Retro orbital pain is caused due to the involvement of trigeminal ganglion (Gessarian ganglion) at the level of Meckel's cave.

Clinical features:

1. Intense head ache (most commonly retro orbital pain)

2. Discharging ear
3. 6th nerve palsy & diplopia
4. Horner's syndrome rarely if sympathetic plexus around internal carotid artery is involved at the level of petrous apex

Pain could be caused by any of the following mechanisms:

1. Referred otalgia
2. Dural irritation in the tegmen area
3. Inflammation of Gessarian ganglion
4. Localized meningitis

It is clearly not known how long it takes for infection to spread from mastoid air cells to petrous apex. Studies have shown that this interval can vary between 1 week to 3 months. The spread of infection from mastoid air cell system to petrous apex depends on the following factors:

1. Type and virulence of the infecting organism
2. Host immunity
3. Pneumatization of petrous apex area – If this area is not pneumatized then infection from mastoid air cell system cannot spread to this critical area.
4. Children are commonly involved because the common infecting organisms in them is H. Influenza which is known to spread rapidly.

Role of Imaging:

High resolution CT scan and MRI studies help in clinching the diagnosis. CT scan reveals clouding of mastoid and petrous air cells. MRI is useful in patients with suspected lateral sinus thrombophlebitis which may be an associated condition in these patients.

Complications:

The proximity of various venous sinuses to the petrous apex has been attributed to be the cause for various complications following Gradenigo syndrome. These complications include:

1. Thrombosis involving various venous sinuses
2. Meningitis
3. Epidural abscess
4. Brain abscess
5. Palsies involving various cranial nerves
6. Horner's syndrome
7. Prevertebral and parapharyngeal abscesses

Management:

Intravenous broad spectrum antibiotics should be started immediately. If there is associated lateral thrombophlebitis then anticoagulants should be considered. After a week of antibiotic therapy if the patient does not show any signs of recovery then mastoidectomy should be resorted to. In children with gradenigo syndrome with associated lateral sinus thrombophlebitis surgery should be resorted to at the earliest.

Intravenous antibiotic regimen:

1. Vancomycin 60 mg /kg/day – 10 days
2. Cefotaxime 275 mg / kg / day – 7 days

Role of steroids:

Injection dexamethazone has been administered in these patients during acute phase in parenteral dose of 0.8 mg/kg/day. This dose ofcourse should be tapered.

Acute pain can be best managed by use of anti-inflammatory drugs.

Citation:

1.
Gradenigo syndrome by drtbalu [Internet]. [cited 2012 Jan 1];Available from:
<http://drtbalu.co.in/gradenigo.html>